COARCTATION OF THE AORTA

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Since the introduction of radical surgical treatment for coarctation of the aortic arch by Crafoord and Nylin (1945) and by Gross (1945), the operation has been widely performed and in experienced hands the mortality of the operation has been reduced to below 10%.

We have carried out a survey of 52 patients (34 males, 18 female) with coarctation who have been under the care of one or other of us, paying particular attention to the natural history of the disease and to the effect of resection upon its various features in 40 of them.

The Clinical Series

The patients' ages ranged from 1 to 55 years (Tables I and II). Of the 40 patients who had resection (see also Diagram) all were aged under 40, and no fewer than 25 were 15 years or older. Of the 12 patients who did not have surgery, 6 were aged over 40 when last seen, but 3 of these have died, the oldest at the age of 55 (Table I).

Mode of Discovery

It is well known that coarctation is often remarkably symptomless (Lewis, 1933; Bramwell, 1947), and 43 of our cases were diagnosed during medical examinations carried out for reasons unrelated to the coarctation. Nine patients presented with symptoms referable to the coarctation. In two of these symptoms were due to dissecting aneurysm of the aorta. Two others had general symptoms due to subacute bacterial infection, and another presented with nephritis (probably focal-embolic, as vegetations were later found at the site of

TABLE I.—*Twelve Patients with Coarctation of the Aorta who did not Come to Resection*

Case	Sex	When Last Seen			Death	
		Age	B.P.	State	Age	Cause
A	F	10	145/80	Well		
B	M	31	180/100	Blackouts. Physically fit		
C	M	31	168/80	Well		
D	F	46	255/140	5-para. Slight dyspnoea		
E	F	46	200/100	4-para. Well		
F	M	40	160/100	Congestive failure. Auricular fibrillation. Aortic stenosis and incompetence. Mitral stenosis	41	Congestive heart failure. No P.M.
G	F	30	220/130	2-para. Well		
H	M	25	160/90	Heavy manual work. Well		
I	M	42	205/100	Easily fatigued.		
J	M	32	190/110	Physically fit		
K	M	55	170/60	Aortic incompetence and stenosis	55	Congestive heart failure. No P.M.
L	M	47	(80/40, in shock)		47	Dissecting aneurysm of aorta

TABLE II.—*40 Patients with Coarctation of the Aorta who Came to Resection (see also Diagram)*

Case No.	Sex and Age	Date of Operation	Average B.P. before Operation	Diameter of Anastomosis Relative to Aorta Above	Average B.P. after Operation	Femoral B.P. after Operation (Not Average)
1	M 17	1948	144/104		138/82	140/100
2	M 30	1949	237/109		180/103	200/120
3	F 27	1949	210/128		163/95	150/120
4	F 19	1949	187/97		168/90	—
5	F 30	1949	190/133		—	—
6	F 18	1949	190/120		—	—
7	F 14	1950	190/130		175/100	170/120
8	M 13	1951	165/105		138/75	160/100
9	M 6	1951	165/95		115/83	120/?
10	F 33	1952	203/121	45%	170/90	190/?
11	M 25	1952	183/110		162/98	200/130
12	M 20	1952	200/81	60%	177/78	—
13*	M 7	1952	140/90		118/76	120/90
14	M 20	1953	181/90	90%	165/75	150/110
15	M 6	1953	152/93	75%	138/95	115/?
16	M 20	1953	172/105		150/90	140/?
17	M 5	1953	134/98	86%	133/90	140/?
18	M 10	1953	177/100		125/80	155/100
19*	F 36	1953	170/105		—	—
20	F 18	1953	200/90		166/70	160/90
21	M 20	1954	180/90	86%	145/85	145/110
22	M 17	1953	203/111	100%	145/82	—
23	M 5	1954	180/110	84%	155/92	—
24	M 9	1954	208/125	80%	130/72	130/80
25	F 25	1954	186/118	72%	141/86	160/115
26*	M 20	1954	160/95	100%	136/90	160/85
27	F 1	1953	125/80	84%	118/88	115/90
28	M 31	1954	200/100	80%	180/100	—
29	F 7	1954	155/88	100%	140/84	150/90
30	F 3	1954	170/108	100%	112/83	135/75
31	M 5	1954	158/64	100%	108/65	100/60
32	M 7	1954	173/105	92%	124/80	120/85
33	M 35	1954	195/105	85%	165/100	170/140?
34	M 31	1955	170/124	85%	135/90	—
35	M 18	1955	185/120	92%	175/100	180/120
36	F 32	1955	205/105	100%	145/90	—
37	M 32	1955	189/98	100%	176/100	180/110
38	M 19	1955	150/108	100%	135/105	135/100
39	M 6	1955	135/80	95%	110/75	120/70
40	M 23	1955	155/70	100%	147/86	160/110

In all cases the operative procedure was excision with end-to-end anastomosis, except where grafts are indicated.

Average reduction of blood pressure = 32/16 (excluding Case 40—too recent).

* Freeze-dried aortic homograft inserted.

Note.—(1) The brachial blood-pressure figures are average values. The femorals are simple measurements, usually at last visit. (2) The size of the anastomosis is compared with that of the aorta between the anastomosis and the subclavian artery and presented as a percentage of the latter. No figure is given in cases in which detailed measurements were not made.

coarctation). Three developed cerebrovascular accidents; and, finally, one patient with auricular fibrillation, mitral stenosis, and aortic incompetence was admitted in congestive cardiac failure.

The presenting symptoms of these nine patients, who had been previously well, were manifestly due to serious complications. Equally important complications, however, occurred silently in other cases and were unexpectedly found at operation. The dramatic onset of these complications is illustrated particularly by those who suffered vascular accidents. A labourer aged 47, for example, enjoyed perfect health until his sudden collapse at work from a dissecting aneurysm, which proved fatal in a few days.

Discovery of Symptomless Cases

In several instances the cardiovascular system became suspect many years before the correct diagnosis was reached. Four patients, for example, had been diagnosed in childhood as having rheumatic heart disease on discovery of a cardiac murmur. In fact, an unexpectedly high incidence of rheumatic fever has been reported in patients with coarctation, and three of our cases gave histories of it. One further patient had the signs of valvular disease, possibly rheumatic, with mitral stenosis, aortic incompetence, and auricular fibrillation, in addition to coarctation.

The initial findings that eventually led to the diagnosis of coarctation in these 43 patients were heart murmurs

Diagram showing anatomy of coarctation in 40 cases resected. The dotted lines indicate site of resection.

in 26, hypertension in 12, pulsating swelling in the neck in 2, and x-ray examination in 3.

The majority of cases were therefore accidentally discovered in the first instance and had no symptoms due to the coarctation. Some had an impressive capacity for hard manual work, enjoyed excellent health, and were object-lessons of how little incapacity coarctation of itself need cause. Three patients who have not had resection and are now over 40 are conspicuous examples:

Case I, a steel-bender, was advised at the age of 38 (B.P. 200/100) to change his job for a lighter one. At 42 (B.P. 205/100), he self-consciously confessed that he had not found it necessary to take this advice.

Case D had a blood pressure of 220/110 at 28 and was advised about her heart. She next came for advice during her fifth pregnancy, was with difficulty persuaded to relinquish her part-time job as a porter, and went intrepidly to term with a normal delivery. At 46, and grossly obese, she admitted to a little dyspnoea (B.P. 250/125).

Case E led an active life and has normal health at the age of 46 (B.P. 200/100) (B.P. 170/100 aged 28). She had four full-term deliveries (Table I).

Intercurrent Symptoms

As a result of one or other of the initial findings, 23 patients had had attention drawn to their hearts a consider-

able time before coming under our observation, and by then were 18 years of age or older. Eighteen had developed one or more of the following symptoms: headaches, dizziness, palpitations, dyspnoea of effort, easy fatigability, faints, precordial or other chest pain, and pains or heaviness in the legs. There were no symptoms in five cases.

By contrast, four other patients attained the age of 18 years or more without being aware of any cardiovascular abnormality until coarctation was discovered. All were symptom-free.

This contrast alone suggests that many of the symptoms which developed in the 18 patients were due to cardiac anxiety. The clinical assessment of their symptoms, though often difficult for this very reason, led us to the same general conclusion. The correspondence between the list of symptoms that had developed under awareness of a cardiovascular defect and that noted by Wood (1941) in Da Costa's syndrome is further evidence of their neurasthenic nature. Again, eight asymptomatic patients were observed by us for a year or more (mean period two to five years) between the making of the diagnosis and the performance of the operation; during this period of observation three developed symptoms due to a mild cardiac anxiety state despite attempts at prevention by assurance and explanation.

It is unnecessary to attribute this disconcerting incidence of anxiety neurosis to any special liability of subjects with coarctation to develop it; it is a well-recognized consequence

of the diagnosis of heart disease in any form. White (1951) found an incidence of neurocirculatory asthenia (effort syndrome) of 4.7% in cases of organic heart disease. In only two of our cases was neurosis incapacitating, and these had very inadequate personalities; however, even patients of ordinary stability of mind showed a tendency to develop anxieties about their health to a degree which was sometimes sufficient to encroach on their well-being and working capacity.

Fainting attacks occurred in eight patients; they were usually multiple, but were single in one instance. No clear cause could be found in any, but one patient developed a spontaneous subarachnoid haemorrhage later. The attacks were not observed by us, but appear to have been a simple syncope, usually with loss of consciousness for a few moments. It is of interest that in all of these patients attention had been called to the cardiovascular system before the attacks began, and it will be realized that Wood (1941) reported syncope in 34.5% of his cases of Da Costa's syndrome. However, from the lack of corroborative evidence of psychoneurosis in some cases we were reluctant to ascribe all the episodes of syncope to this cause, although unable to provide an alternative explanation.

Pain in the front of the chest was experienced by five patients. It never had the typical features of angina pectoris and was usually rather nondescript. In two patients it had some characteristics of intercostal neuralgia, and it is possible that pressure from the dilated anastomotic intercostal arteries was responsible. No neurological signs of spinal cord or root affection were found in these or our other patients, although such have been described (Wyburn-Mason, 1943).

Pain in the legs could not with certainty be ascribed to ischaemia and was not typical of intermittent claudication, but consisted rather of aching or heaviness after effort, or towards the end of the day. The symptom occurred in eight patients, all of whom had been aware of a cardiac abnormality for many years.

Although many of the symptoms that develop in persons with coarctation may have no organic basis, serious mischief may proceed silently under their cover.

A man (Case 26) aged 20 complained of tiredness, dyspnoea, vague pains in the feet, and headaches for about a year, during which time no apparent cause was discovered and psychiatric treatment was given. Eventually embolism of the dorsalis pedis artery disclosed the true state of affairs, and on transfer to Hammersmith Hospital blood cultures confirmed the diagnosis of subacute bacterial infection due to *Streptococcus viridans* associated with a coarctation.

Epistaxis

Epistaxis occurred a number of times in five patients. One of these had a cerebrovascular accident later; in his case and in three of the others the blood pressure was only moderately elevated; in the fifth it was unusually high.

Complications

The complications which were found either clinically or at operation are described below.

Subacute Bacterial Infection

One patient (Case 26) had manifest subacute bacterial endocarditis or endoaortitis when first seen by us.

Case 26.—The "functional" nature of his symptoms had led to their being attributed to neurosis until subacute bacterial infection declared itself by an embolus in the dorsalis pedis artery. Culture of *Str. viridans* from the blood confirmed this diagnosis. The infection responded to penicillin before operation. Thoracotomy revealed a post-stenotic saccular aneurysm of the aorta, 45 mm. in diameter. Six centimetres of the aorta, including the aneurysm, was resected, being replaced by a freeze-dried aortic homograft. The resected specimen showed a coarctation with a 2-mm. orifice opening into a dilated aorta, from one wall of which the aneurysm bulged. At the site of the aneurysm the media was totally interrupted and replaced by fibrous tissue;

vegetations showing inflammatory cell infiltration were found on the intima (Fig. 1). A year after operation this patient remained perfectly fit.

Vegetations were found on the resected segment of aorta in seven further cases: in four of these no suspicion of infective endoaortitis had been entertained.

Case 14.—A young man had been under observation for two years before operation, yet, though vegetations on the aortic intima were found just below the coarctation from which *Str. viridans* was cultured, nothing could be discovered other than a period of low-grade fever—99° F. (37.2° C.)—on looking back through his records, which would enable the diagnosis of endoaortitis to be made. After operation he was treated with penicillin for six weeks. Neither then nor during the follow-up of over two years has there been any evidence of further infection.

Case 17.—The second silent case was that of a boy whose resected segment of aorta contained vegetations, sterile on culture, but otherwise resembling, both grossly and microscopically, those of the preceding case. Looking back, occasional evening temperatures of 99° F. (37.2° C.) had been recorded, though nothing more suggestive than this was discovered. The blood sedimentation rate was 3 mm. in one hour (Westergren).

Case 12.—The third case was that of a young man of 20 years who was symptom-free and had no signs of infection. Sterile vegetations were found at the site of coarctation. Both this patient and Case 17 remained well for over two years from the operation.

Case 40.—In the fourth case there was no clinical evidence of infection. Unusually exuberant vegetations were found arising from the coarctation. Macroscopically they resembled those just described, but microscopically were found to consist of almost acellular fibrin.

Three further cases (Nos. 18, 29, and 32) had fever during pre-operative hospital observation. There were no signs of infective endoaortitis and blood cultures were negative, but, as experience had shown how insidious the process could be, antibiotics were given before operation. In each case sterile vegetations were found on the aortic intima. All three patients have remained well since operation.

Case 18.—This patient was a boy in whom a cardiac abnormality was diagnosed when he was admitted to hospital because of nephritis. His low fever, never beyond 100° F. (37.8° C.), was unaccompanied by symptoms. He was treated with penicillin for three weeks before operation. A plaque of adherent fibrous peri-aortitis was found on the medial aspect of the aorta, 5 mm. below the coarctation. It measured 1 cm. in diameter and overlaid a patch of intimal vegetations which extended from the mouth of the stenosis on to the aorta just below. The vegetations were sterile on culture, and the histological appearances were uninformative regarding infective aetiology.

In eight cases, therefore, vegetations were encountered on the aortic intima at and below the coarctation, and in only one was infective endoaortitis clinically evident.

The infective nature of these vegetations is not certain except in Cases 14 and 26, in which *Str. viridans* was cultured from the vegetations and the blood respectively and clumps of bacteria were seen in the histological sections. Such bacteriological evidence was not to be expected in the four others who had pre-operative antibiotic therapy, but in these, and in two others the appearances of the lesions closely resembled those of Case 14. It is therefore possible that all had an infective origin.

Acellular fibrin deposits on the intima were recognized in several other cases, but their macroscopic and histo-

FIG. 1.—Photograph of resected coarctation and aneurysm in Case 26. The specimen has been bisected longitudinally and laid open to show both halves. The coarctation can be seen at the upper end of the specimen.

logical appearances were distinctly different. These are the so-called "jet lesions" described by Edwards *et al.* (1948).

Among Abbott's (1928) 200 cases of coarctation were 14 instances of infective endoaortitis. Four of these had resulted in rupture of the aorta. The silence of the infection is sinister, therefore, in that disaster may happen before the complication is recognized and before antibiotic therapy is indicated.

Dissecting Aneurysm of the Aorta

The calamity of dissecting aneurysm of the aorta was encountered twice. The sudden onset and the rapidly fatal issue in one case have been mentioned :

Case L.—A labourer (Table I) aged 47, previously in excellent health, collapsed at his work. Comatose on admission to hospital, he remained in coma until he died on the fourth day. Necropsy showed coarctation with a dissecting aneurysm of the

aorta. The initial tear was a short distance above the aortic valve, and thrombus formation here had resulted in embolism of the brain and kidneys before final aortic rupture into the pericardial sac caused death from tamponade.

The second was of slower tempo, and an attempt was made to deal with it surgically.

Case 19.—A woman who had led an active life with good health had tonsillitis in September, 1952 (aged 35), followed three weeks later by severe interscapular pain radiating down the back. (B.P. 170/100). The femoral pulses were feeble and delayed. X-ray examination revealed an aortic aneurysm (Figs. 2 and 3) and a retrograde aortogram showed a dilated aorta with a double lumen. In March, 1953, she had a further attack of interscapular pain. A diagnosis of dissecting aneurysm was made. In October, 1953, thoracotomy revealed coarctation below the origin of the left subclavian artery with dissection of the aorta below, giving entrance to a large aneurysm; there was a double lumen down the entire length of the thoracic aorta extending into the abdominal aorta for an undetermined distance (Fig. 4). The thoracic aorta from the subclavian artery to the diaphragm was resected and replaced by a freeze-dried graft. The specimen showed that the initial tear was 2 cm. below the coarctation. There were no vegetations on the excised specimen. The patient made a good recovery, but in the convalescent home, six weeks after operation, a massive fatal haematemesis occurred. Necropsy revealed that the dissecting aneurysm had extended throughout the abdominal aorta to the bifurcation and had ruptured into the stomach. There was gross mucoid change of the media with corresponding loss of muscle in the aorta proximal to the graft and in the subclavian artery.

The operation was a desperate expedient in the hope that success might attend replacement with a graft if the dissection was favourably limited. This type of dissecting aneurysm must be regarded as one of the direct complications of coarctation.

Non-dissecting Aortic Aneurysm

Saccular aortic aneurysms were recognized radiologically and found at thoracotomy in two cases. In each the aneurysm lay immediately beyond the coarctation. One of the patients (Case 26) had subacute bacterial infection and has been described; the other (Case 13) did not have an infective lesion.

Case 13.—The diagnosis of coarctation was made by Dr. W. Brigden in this boy aged 7, who had pains in his limbs, thought at first to be rheumatic. X-ray films showed a large shadow just below the aortic arch, and its aneurysmal nature was confirmed by angiocardiology (Fig. 5). At thoracotomy, a saccular aneurysm, 35 mm. in diameter, began 20 mm. below the left subclavian artery. The main mass of the aneurysm extended deeply behind the hilum of the lung, and the wall was quite firmly attached to the spine and firmly blended with the oesophagus. The muscular layer of the oesophagus had been eroded and only oesophageal mucosa covered the thin wall of the aneurysm at this point. The aneurysm was mobilized, excised, and replaced by a 35-mm. aortic graft. In the specimen a coarctation with a 1-mm. orifice led eccentrically into the aneurysm, which had a large distal opening. During the three years of follow-up the patient had remained very well. The angiocardiology, 20 months after resection, showed a normal aorta (Fig. 6).

Such aneurysms can be resected, although their presence adds to the risks of the operation. But the degree of

FIG. 2.—Six-foot (1.8 m.) postero-anterior radiograph of chest in Case 19, showing the aneurysm of the arch and descending aorta.

FIG. 3.—Six-foot (1.8 m.) postero-anterior radiograph of chest in Case 19 after resection of the aneurysm and insertion of aortic graft.

FIG. 4.—Excised aneurysmal sac in Case 19 viewed from below. The double lumen is well shown, the false channel being the larger, above, and the true lumen the smaller, below.

erosion and thinning found in Case 13 indicates that its initial manifestation might easily have been fatal rupture into the oesophagus. Although their tempo is slower than that of dissecting aneurysms, their silence makes their timely discovery so much a matter of chance that they are scarcely less hazardous to life.

Post-stenotic dilatations were common. At operation they added to the difficulties of obtaining a uniform calibre of the reconstituted aorta.

FIG. 5.—Angiocardiogram, left lateral projection in Case 13. The aneurysm is clearly shown in the upper portion of the descending aorta. There is no filling defect within the sac, and the descending aorta appears normal. The coarctation is hidden by the aneurysm. (Film reproduced by courtesy of Dr. Wallace Brigden.)

FIG. 6.—Post-operative angiocardiogram of Case 13: left lateral projection. No aneurysm is present, and there is good filling of the aorta with minimal constriction of the lumen.

Intercostal Aneurysms

Small saccular aneurysms occurred even more frequently on the intercostal arteries just below the coarctation, most commonly close to their origin from the aorta (Fig. 7). They were found at operation in six of our cases (Nos. 10, 20, 25, 34, 36, and 37), and their walls were so delicate that blood could be seen swirling inside them. We were surprised to find no record of their rupture in the literature, but such an event might be mistaken for rupture of the aorta itself. Dr. Norah Schuster (1955, personal communication), who noted them intact in two cases of coarctation at necropsy, was of the opinion that they might very easily be overlooked. Their common incidence at operation and the hazard they constitute are attested by Gross (1953).

These intercostal aneurysms have all been clinically silent. Furthermore, they were not evident on x-ray examination, except in two of our cases where they indented the barium-filled oesophagus (Fig. 8). Angiocardiography also failed to outline them. In short, they are part of the occult threat of fatal vascular rupture that besets persons with coarctation.

FIG. 7.—Photograph of resected aorta in Case 20. A thin-walled saccular aneurysm can be seen at the junction of an intercostal artery with the aorta just below the coarctation on the right-hand side of the picture. The coarctation can be seen at the upper end of the specimen.

FIG. 8.—Chest radiograph with barium swallow (left lateral projection) in Case 10. The upper part of the oesophagus is displaced forward by the post-stenotic dilatation of the aorta immediately below the coarctation. Below this is another more sharply localized indentation of the oesophagus produced by an intercostal aneurysm.

Intracranial Vascular Lesions

Cerebrovascular accidents occurred in three of our patients.

Case 2.—Male. Cardiac abnormality discovered at birth. 1944 (aged 25): complained of frontal headaches. 1947: "stroke" with hemiplegia, with almost complete recovery; coarctation diagnosed. 1949: resection of coarctation; before resection, average brachial blood pressure was 237/109; after resection average brachial pressure, during six years follow-up, was 180/103. No further cerebrovascular accidents.

Case 8.—Male. 1950 (aged 12): sudden onset of headache when diving into swimming-pool; meningism and impaired consciousness followed; brachial blood pressure 168/104; sub-arachnoid haemorrhage confirmed by lumbar puncture; good recovery. 1951: resection of coarctation. Followed up for five years—remained very well. Average brachial blood pressure: pre-operative, 165/105; post-operative, 138/75.

Case 22.—Male. 1953 (aged 17): classical spontaneous sub-arachnoid haemorrhage; slow recovery; resection of coarctation. During two years' post-operative observation remained well. Average brachial blood pressure: pre-operative, 203/111; post-operative, 145/82.

The brachial blood pressure in these three patients was higher relative to their age than in most of our cases, and doubtless was a factor in the aetiology of the intracranial lesions.

Hypertension

In all our cases the brachial blood pressure was elevated for the individual's age and sex above the average value expected in the general population (Stocks and Karn, 1924; Hamilton *et al.*, 1954). The elevation affected both the systolic and the diastolic pressure, except where the latter was lowered by aortic incompetence. The hypertension was usually moderate in degree, and it increased with age after the fashion of the blood pressure in the population generally (Hamilton *et al.*, 1954), but at a somewhat accelerated pace. Rapid rises in pressure or rapid deterioration of the patient from increasing hypertension were observed in only one patient (Case 7), a girl who, whilst under observation, developed papilloedema in association with unusually high levels of blood pressure.

Case 7.—In 1948, when aged 12, this girl was suspected of having rheumatic fever with carditis. Coarctation was discovered, but it was possible that active rheumatism was also present. Later, tuberculous gingivitis and cervical adenitis developed. In November, 1949, she could not see the blackboard at school. Bilateral papilloedema and one superficial retinal haemorrhage were present with arteriovenous nipping. Brachial blood pressure 200/125. Femoral pulses barely palpable, and femoral blood pressure not obtainable. September, 1950, retinopathy had not progressed. Blood pressure 190/130. Clinical urine tests normal. Effective renal plasma flow 278 and 415, and glomerular filtration rate 132 and 157 ml. a minute (duplicate measurements). Resection of coarctation with direct anastomosis carried out. Papilloedema subsided. Remained well during five and a half years' post-operative observation. Albuminuria noted in October, 1953, but not subsequently. *Present assessment:* Condition of optic disks consistent with healed papilloedema; no retinopathy; no albuminuria; renal plasma flow 268 and 344, and glomerular filtration rate 41 and 60 ml. a minute; no delay in femoral pulse peak; no palpable collateral vessels; average brachial blood pressure after operation 175/100 (before operation 190/130). The change in renal clearances suggests deterioration of renal function.

So far as we know, papilloedema has not previously been described in coarctation, and the hypertension existing in the upper parts of the body has not been reported to enter a malignant phase. Case 7 seems to be a unique exception.

The probable part played by hypertension in the production of intracranial vascular lesions in three of our patients has been mentioned, and this point and the aetiological role of hypertension in dissecting aneurysms are more fully discussed below. Apart from this, the hypertension of coarctation was remarkably well tolerated. Uraemia did not occur, and heart failure was found only in association with valvular defects of the heart.

Heart Failure

Congestive heart failure occurred in only two patients, but caused death in both.

Case F.—Male (Table I). Congenital heart disease was diagnosed at school, but he lived an active life until the age of 33, when dyspnoea and recurrent bronchitis began. He continued to work intermittently until aged 40, when nocturnal dyspnoea and oedema of the legs set in. Auricular fibrillation was then present and the heart was enlarged. The first heart sound was accentuated and there was an apical diastolic rumble; a systolic murmur with thrill was present at the base of the heart and an early diastolic murmur along the left sternal border. Blood pressure was 160/100, and the usual signs of coarctation were present. E.C.G. showed left bundle-branch block. He died at home (aged 41) a year later. No necropsy was performed. Clinical diagnosis: coarctation of aorta; aortic incompetence and probably aortic and mitral stenosis.

Case K.—Male. Heart disease known since childhood. There had always been some dyspnoea of effort. Pneumonia 1949 and 1952. On examination (1952, aged 55) gross cardiac enlargement, chiefly left ventricle. Loud systolic murmur at base; early diastolic murmur at left sternal edge; apical mid-diastolic murmur. Blood pressure 170/60. X-ray film showed calcified aortic valve and dilated ascending aorta. E.C.G. left ventricular hypertrophy. Heart failure imminent. Died at home

two months later. No necropsy. Clinical diagnosis—coarctation of aorta; aortic stenosis and incompetence.

Severe valvular lesions were present in both these patients, and presumably accounted for failure of the heart. Hypertension was not severe in either at the time of observation.

Natural History of Coarctation

The high incidence of serious events and ominous operative findings in our patients is in line with what is known of the natural history of coarctation. Necropsy series indicate that the majority of patients with the "adult type" of coarctation die before the fifth decade of life. Seventy-four per cent. of Abbott's (1928) collected cases (200 cases, including none under 3 years of age) and 61% of 104 additional cases collected by Reifstein *et al.* (1947) had not survived the 40th year of life. Such statistics, being derived from necropsies, may give an unduly gloomy impression; but, unfortunately, available clinical studies which should give a truer perspective are much less extensive.

Lewis (1933), Bramwell (1947), Campbell and Suzman (1947), Newman (1948), and Shapiro (1949) give clinical accounts of a total of 94 cases of coarctation, of which 27 were fatal. When death was from cardiovascular causes (excluding two patients who died after surgical operation), the average age at death was 33 years, and only seven had passed the 40th year. Where the ages of the remaining subjects were given, only six were recorded to have passed the 40th year of life.

The evidence is clear that many if not most patients with coarctation die before middle age.

Causes of Death

Although they may have no symptoms persons with coarctation live in danger. Vascular complications are often sudden and overwhelming, and may preclude effective treatment when they have set in; they challenge resection to prove itself prophylactic as well as anatomically corrective. The vascular complications are sometimes remote from the coarctation, and analysis of the mode of death, in its relationship to coarctation as the basic lesion, is of great importance.

The region of the coarctation itself is sometimes the seat of fatal lesions. In our cases, actually or potentially infective vegetations were found eight times. Excision of the coarctation should abolish the risks from this complication. However, infective endocarditis is more common than infective endoaortitis in coarctation. Of the 23 cases of one or other in the series of Reifstein *et al.* (1947), only six had endoaortitis without endocarditis, and the large majority of cases of the latter lesion had infection of bicuspid aortic valves. Since the same authors found bicuspid aortic valves in 42% of all cases of coarctation, it is obvious that a considerable part of the hazard from infection resides in this lesion alone, which would not be expected to be diminished by resection of the coarctation.

The other local lesion in reference to coarctation is aneurysm formation, which was found twice in the post-stenotic aorta and six times in the intercostals entering below the stenosis in our cases. One further case had a dissecting aneurysm with the initial tear just below the coarctation. In 5 of the 104 cases of Reifstein *et al.* (1947) rupture of the aorta had taken place beyond the coarctation. Abbott (1928) also found rupture in direct relation to the stricture in 5 of her 200 cases. Prevention of this complication may also reasonably be expected from resection and reconstitution of a full lumen.

Other vascular lesions are more remote from the coarctation. The initial tear in our other patient with dissecting aneurysm was located just above the aortic valve. It was situated in the ascending aorta in 19 of the cases of Reifstein *et al.* and in 33 of those of Abbott. The aortic valve was bicuspid in no fewer than 17 of the 33 cases reported by Abbott, and it is noteworthy that Gore (1953) found bicuspid aortic valves in 9 out of 32 cases of dissecting aneurysm in persons aged under 40 (in these it was asso-

ciated with coarctation only once). The initial tear was in the ascending aorta in the great majority. By contrast with coarctation Gore found that hypertension was exceptional in cases of dissection under the age of 40 though the rule after that age, and concluded that the elevated blood pressure exploits the weakness of primarily diseased or abnormal vessels, though it might also aggravate the degeneration of the media which underlies dissection.

Intracranial vascular lesions occurred in three of our cases. Subarachnoid haemorrhage was proved in one. The high incidence of intracranial aneurysms of the congenital type is well recognized in coarctation (Abbott, 1928), and rupture of these appears to constitute the main hazard from intracranial lesions, though cerebral thrombosis may also occur. Reifstein *et al.* (1947), reporting 11 cases of intracranial lesions among their 104 necropsied cases, found no instance of cerebral haemorrhage that had not originated in an aneurysm, either "congenital" or mycotic. Hypertension is provocative of rupture of these aneurysms, but rupture may occur also in normotensive persons (Taylor and Whitfield, 1936; Magee, 1943; Walton, 1952; Stehbens, 1954).

Hypertensive heart failure did not occur in our cases, although hypertension, by adding to the load placed on the left ventricle by aortic valve disease, may well aggravate failure. The great majority of the patients of Reifstein *et al.* and of Abbott who had heart failure also had other cardiac lesions, but Abbott remarked that there was a relatively small group in which coarctation and the secondary changes in the circulation that arise from it had apparently been the essential factors in the cardiac strain. It is in early childhood, possibly before the collateral circulation is well developed, that the heart most often fails from the hypertension of coarctation (Mustard *et al.*, 1955), although even at this time there are often associated lesions such as persistent ductus arteriosus and endocardial fibro-elastosis. Reduction of the blood pressure can be expected to prevent some of the complications that occur at a distance from the coarctation, and this reduction is one of the chief aims of resection.

Pregnancy

The risk of rupture of the aorta in patients with coarctation is commonly believed to be increased by pregnancy. Mendelson (1940), reviewing the literature, cited 31 cases of pregnancy in patients with coarctation; dissecting aneurysm had occurred in two patients. Ten of our patients had one or more pregnancies, and their obstetrical histories were of interest. Three of these patients did not have surgical treatment; one had had five and another had had four full-term deliveries without incident; the third patient had had two pregnancies, both terminated at term by elective caesarean section. Six patients had had pregnancies before being referred for surgery—two had had one pregnancy; three had had two pregnancies, and one had had three full-term normal deliveries before resection.

The indications for therapeutic abortion were the fear of rupture of the aorta rather than actual deterioration of the patient. Blood-pressure measurements showed a lowering of the pressures in the middle trimester; there was usually a rise of pressure in late pregnancy and during parturition. This corresponds with the findings of Dixon and Hartley (1955).

Our experience of pregnancy in coarctation is too slight to permit conclusions of a general character; but clearly, while great care in management is required, pregnancy can be brought to a successful conclusion in many cases. Two of our patients have had normal pregnancies and deliveries following resection of the aorta.

Physical Signs

Some of the most important signs in the assessment of patients may be mentioned.

Habitus.—The development of stature was retarded in many cases. The lower limbs were underdeveloped, the

pubis-to-ground measurement was short, whilst the upper parts of the body were particularly well developed and sturdy, and span exceeded height—in one case by as much as 5 in. (12.5 cm.). Disproportion was most pronounced in two patients in whom the aorta was completely interrupted by an imperforate diaphragm.

"Carotid Swell."—The augmented carotid pulsation was very characteristic, and of itself suggested the diagnosis. A sustained systolic arterial wave or "swell" appears in the suprasternal fossa and travels outwards along the subclavian arteries and up the carotids, where it is best observed.

Collateral Arteries.—These were most often found around the scapulae, but examination of the intercostal spaces, the axillae, and the posterior triangle of the neck sometimes revealed them. Such enlarged anastomotic arteries were found in 42 of our cases. They were absent in 10 patients, whose ages were 3, 5 (two cases), 7, 10, 18, 19, 25, 30, and 36 years. Thus their absence was not explicable by youth of the patients, nor did it correlate with the disproportion in habitus.

Murmurs

Systolic murmurs are a feature of coarctation. The precordial systolic murmur was often nondescript, but a characteristic murmur occurred in many cases: loudest at the sternal borders, its onset separated from the first heart sound by a brief silent interval, it slowly reached maximum intensity and declined through the second heart sound into early diastole. Distinct from this, in nine cases which came to surgery there was a loud rough systolic murmur at the base, accompanied by a basal systolic thrill in six. In the other three, a thrill was not palpable over the praecordium but only over the carotid arteries. These murmurs suggested aortic stenosis, but for lack of corroboratory signs in the other seven this diagnosis was made in only two cases. The systolic gradient across the aortic valve was measured in these two cases; in one it was 20 mm. and in the other 10 mm. These gradients suggested mild stenosis, and aortic valvotomy was not attempted. Post-operatively, their murmurs were unchanged. Of the remaining seven cases, one came to necropsy and the aortic valve was bicuspid with somewhat thickened leaflets; three others had no murmurs at the base of the heart after operation, and three had fainter murmurs without thrills. It will be recalled that in one patient, who did not have surgery, a clinical diagnosis of aortic stenosis was made and calcification of the aortic valve was seen on x-ray examination.

Subaortic stenosis is known to occur in association with coarctation (6% of Abbott's cases), but our experience here indicates caution in making this diagnosis from the precordial signs alone. From the post-mortem findings in one case it is tempting to ascribe these rough basal systolic murmurs to bicuspid aortic valves, but their diminution after operation seems to indicate that they had included a component from the coarctation or the collateral arteries. We would therefore prefer to underline the difficulty of their interpretation (Smith and Matthews, 1955). One patient (Case 27) had a loud pansystolic murmur maximal in the third left intercostal space, thought to be due to a ventricular septal defect.

Over the upper dorsal spine, and sometimes widely conducted up and down, there was a prolonged murmur, again of late onset with reference to the first heart sound and continuing well into cardiac diastole. A murmur of similar character and timing could occasionally be traced to its origin in a superficial anastomotic artery and be caused to disappear by digital occlusion of this vessel.

The delayed dorsal interscapular systolic murmur may arise in the coarctation itself or in dilated and tortuous intercostal and anastomotic vessels. In one instance a thrill felt at operation on a patient with a complete stenosis appeared to be caused by blood entering the distal aorta from the intercostal arteries. These murmurs are heard in the majority, and their site is a rough guide to the site of the coarctation. When the coarctation is at the usual site the murmur is loudest between the scapulae in the

region of the third dorsal spine. By contrast, however, in lower thoracic and abdominal coarctations the murmur is best heard in the vicinity of the stenosis, as indicated in the following case (not included in the series).

A married woman aged 26 presented with severe hypertension (B.P. 250/140) in the upper limbs and delayed femoral pulses. Auscultation revealed a systolic and an early diastolic murmur in the epigastrium and upper lumbar region, maximal over the lumbar spine. Slight collateral pulsation to the right of the sternum and in the left scapular region was discovered. An aortogram revealed a coarctation at the level of the renal arteries.

A *mid-diastolic murmur* was heard pre-operatively at the cardiac apex in 15 of the 40 resected cases. It resembled the diastolic murmur of mitral stenosis sometimes very closely, but more often it was shorter and less rumbling, and was inconstant, being clearly audible on some occasions and not at all on others. In 11 of these cases there was radiological evidence of left ventricular enlargement, but this was absent in four. Furthermore, left ventricular enlargement was found in 18 cases in which a mid-diastolic murmur was never heard. In 6 of the 14 cases an early diastolic diminuendo murmur was audible at the left sternal border, but this was also heard in two cases that never exhibited an apical diastolic murmur. Two of the six cases that had both early and mid-diastolic murmurs showed resistency of the ductus arteriosus, and after resection of the coarctation and ligation of the ductus neither murmur could be heard; in the remaining four cases the early diastolic murmur was attributed to aortic regurgitation.

The mechanism of the mid-diastolic murmur is unknown. It was not related to size of the left ventricle, but was present in six of the eight cases with aortic regurgitation, although the converse did not hold. The cases associated with persistent ductus are of additional interest in that a similar mid-diastolic murmur has been recognized in that condition without coarctation.

No evidence of mitral stenosis existed in these cases, although amongst the unresected cases was one with heart failure and good clinical evidence of mitral stenosis, but necropsy was not carried out. Minor degrees of mitral stenosis were therefore not excluded, but were improbable.

During post-operative observation the apical mid-diastolic murmur was not heard in six cases where it had been heard before, but the significance of this is open to doubt because of the intermittency of the murmur. Two of these cases had had a persistent ductus arteriosus. The murmur was never heard *de novo* post-operatively. Where it disappeared, the left ventricular size was smaller on x-ray examination in one case but unchanged in the others; the fall in blood pressure was greater than the group mean fall in two cases and less in one. With the exception of the cases with persistent ductus, no reason was apparent, therefore, for the disappearance or continuance of this apical murmur.

An *early diastolic murmur* was heard pre-operatively at the left sternal border in eight resected cases. In all of them, including two cases with wide patency of the ductus arteriosus, it had the usual characteristics of the murmur of aortic regurgitation, and the typical continuous murmur was not heard in any instance. The ductus entered the narrow segment of aorta just above the constriction. The combination of persistent ductus and coarctation must therefore be regarded as another cause of the replacement of the classical continuous murmur of the ductus by separate systolic and diastolic elements. In the resected cases the degree of aortic incompetence was usually slight and never very gross. In two unresected cases, however, gross aortic incompetence was present; both had heart failure and have been described above.

After operation the early diastolic murmur disappeared in two cases of persistent ductus. In one of the cases of aortic incompetence it became louder after operation; this was in a patient who had vegetations on the resected aorta. An early diastolic murmur was heard for the first time during post-operative observation in two cases; in one it

became quite loud, but in the other it was so soft that it may have been missed before. In neither was there evidence of infective endocarditis.

Femoral Blood Pressure and Pulse

The femoral blood pressure was measured with a suitably sized thigh cuff by auscultation over the popliteal fossa, and by palpation of the pulse at the ankle where possible. In this connexion it should be emphasized that the posterior tibial and dorsalis pedis pulses are sometimes palpable in coarctation even when the volume of the femoral pulse is appreciably diminished. In all cases the femoral pressure was either unrecordable or was of lower systolic value than the brachial pressure measured at the same time. The femoral diastolic pressure is difficult to measure with accuracy by this method, but when readings were obtained the values approximated to the brachial diastolic pressure. Normal values for the femoral blood pressure in large samples of the population are not available, but Steele and Cohn (1938) investigated it by arterial puncture in healthy subjects and found that it approximated to the radial pressure.

In all cases the femoral pulse wave was either of small amplitude with delayed peak or could not be felt. The delay of the pulse wave peak, well described by Lewis (1933), and a cardinal sign of coarctation, can be appreciated clinically by simultaneous palpation of radial and femoral pulses. In health the femoral pulse peak is reached before or synchronously with the radial, but coarctation delays the wave so that the femoral peak is reached later than the radial. Graphic registration of the radial and femoral pulses is of assistance when clinical doubt exists (Fig. 9) (Counihan, 1956).

Cardiography

Cardiograms were available in 48 patients; they showed the pattern of left ventricular hypertrophy in 26 and suggested it in 6 others. No evidence of left ventricular hypertrophy was seen in 12 cases. Right bundle-branch block was found in two and left bundle-branch block in one case—

FIG. 9.—Femoral and radial artery sphygmograms and simultaneous cardiograms before and after operation in Case 7. The delay in the pre-operative femoral pulse is clearly shown, the peak occurring at the summit of the T wave of the cardiogram as compared with the peak of the radial wave, which occurs at the onset of the T wave. The post-operative tracings show that the delay has been abolished. [Reproduced by kind permission of the Editor of *Clinical Science*.]

this was the case of heart failure with aortic and mitral valve disease, and the only instance of auricular fibrillation. One patient (Case 40) had the Wolff-Parkinson-White syndrome.

The cardiographic position of the heart is more commonly vertical in coarctation than in other hypertensive states. Among our cases a vertical position or intermediate position was found in the majority. In one case (No. 20) Lead V_4R showing an rsR pattern, suggesting right ventricular hypertrophy in addition to left (Camerini *et al.*, 1955).

Radiology

X-ray examination of the chest provides further signs of coarctation and helps to reveal its size and the state of the aorta. The x-ray films of 40 cases were available for this study.

Deformity of the aortic arch is an important radiological sign. The arch was abnormal in 27 cases—large or aneurysmal in 4, absent in 8, small in 5, and double in 10. The arch appeared normal in the remaining 13 cases in the postero-anterior view.

Dilatation of the ascending aorta was found in 15 cases, and was of aneurysmal proportions in one. The descending aorta displaced the barium-filled oesophagus forward in 22 cases. This valuable sign, best seen in the left anterior oblique view (Fig. 8), was present in 6 of the 13 cases in which the aortic arch appeared otherwise normal, and raised the number of cases to 33 in which radiology without vascular contrast media permitted location of the site of coarctation in the aortic arch. The forward kink of the aorta at the point of coarctation and post-stenotic dilatation caused this displacement of the oesophagus. Post-stenotic dilatation had reached aneurysmal proportions in two cases. Also in two cases aneurysm of a post-stenotic intercostal artery produced a shorter indentation of the oesophagus; in one of these the intercostal impression was in addition to that produced by the descending aorta (Fig. 8).

Notching of the lower rib margins from dilated tortuous intercostal arteries was found in the x-ray films of 31 cases. In five instances rib-notching was present on one side only; the right side was involved on four occasions, and the left once. Unilateral rib-notching may be caused by an abnormal origin of the left subclavian artery which arises from or near the coarcted segment, but such an anomaly accounted for only one of our cases, whilst a second patient with an abnormal subclavian had bilateral rib-notching. Greatly enlarged internal mammary arteries were occasionally identifiable as comma-shaped shadows behind the manubrium sterni in lateral films.

Left ventricular enlargement was seen on x-ray examination in 30 cases. The left ventricle appeared normal in size in nine, and was questionably enlarged in one. Great enlargement of the heart was found only when gross valve lesions were present, and when encountered should suggest a complicated coarctation.

Angiocardiology

Angiocardigrams were made through the venous route in 14 cases, according to the method described by Goodwin *et al.* (1953), and the site of coarctation was outlined in all of them. The origin of the left subclavian artery was well defined in 13, and the estimated distance from it to the proximal end of the coarctation corresponded well with the actual length found at operation. Measurement of the diameter of the aorta proximal to the coarctation on the angiogram also corresponded well with that found at operation.

Venous angiography often overestimated the length of the coarctation and the diameter of the distal aorta. As a result, prediction from angiograms that grafts would probably be necessary was open to error. In four cases the angiocardiology suggested a long narrow coarctation which was found at operation to be a stricture suitable for primary anastomosis. Even in retrospect, angiocardiology did not reveal any of the intercostal aneurysms.

Aortograms were made in two cases by catheterizing the radial artery and introducing the tip of the catheter into the aortic arch (Brodén *et al.*, 1948; Jönsson *et al.*, 1948). An excellent outline of the coarctation was obtained, surpassing in definition the angiograms by the venous route.

When aortic homografts or substitutes are available, we believe that angiography is unnecessary unless atypical features are present. When grafts are not available, however, angiography is advisable. If exact anatomical detail is demanded, aortography supplies it better than venous angiocardiology.

The Surgical Operation

Forty of our cases had a resection of the coarctation. Full technical details of the operation are beyond the scope of this paper, but certain points of interest and importance are worthy of record.

Anaesthesia.—This has been with thiopentone, curare, nitrous oxide, and oxygen. The hypotensive agent trimetaphan camphorsulphonate ("arfonad") has been employed in the last 15 cases and has made mobilization of the aorta and the subsequent anastomosis a great deal easier. There have been no instances of an unusual rise in blood pressure after operation following its use.

Technique

In every case the aim has been to perform an end-to-end anastomosis that achieves a full aortic lumen at the site of the union. In most cases the diameter at the site of anastomosis was 80% or more than that of the aorta immediately above it. In 25 cases where accurate measurements were made, 9 had an anastomotic diameter equal to that of the aorta above (100%), and 12 had diameters between 80% and 100%. In only four was it less than 80% (Table II).

Measurements of the diameters were made with adjustable callipers and provided an estimate of the size of the lumen of the aorta. In general, the diameter of the aortic segment beyond the left subclavian artery but proximal to the coarctation approximates the diameter of most of the descending aorta in the chest. Comparison between this diameter and that at the site of the anastomosis therefore afforded a method of measuring the success attained in achieving a uniform aortic lumen, and in restoring normality. Because of the frequency of post-stenotic dilatation, the diameter of the aorta just beyond the coarctation was a less accurate guide. Comparison of diameters was not made when the proximal segment was very distorted, and was out of the question when the coarctation abutted directly on the origin of the left subclavian artery.

The anastomoses have been done with 000 silk, using a continuous everting layer posteriorly and interrupted everting mattress sutures anteriorly. Originally 00000 silk was used, but the stronger sutures were employed after our experience in Case 6, when the suture line gave way six hours after operation owing to an actual break in the continuous posterior layer of silk. The use of a modified Broem's vice has made the anastomosis a much simpler and safer procedure, as the divided ends of the aorta can be maintained in constant relationship and both clamps can be moved as a unit.

Freeze-dried aortic homografts have been available for the last 20 cases, but it has been our policy to reserve their use for cases with very definite indications until more is known about their ultimate fate. We have found it essential to use a graft in only two cases of post-stenotic aneurysm and one case of dissecting aneurysm. Only two of the earlier cases (Nos. 10 and 12) would, in retrospect, have benefited from a graft if one had been available. One (Case 10) was a patient with a diaphragmatic type of coarctation with 4 cm. of hypoplastic aorta distal to the coarctation. As much of the hypoplastic aorta as possible was resected. The diameter of the aorta at the site of anastomosis was 9 mm. and that of the aorta immediately above was 20 mm.

Operative Findings

In all cases the coarctation was of the classical "adult" type. Dissecting aneurysm of the aorta was present in one case, non-dissecting post-stenotic aneurysm of the aorta in two cases, and berry aneurysms of the intercostals in six cases. These last usually lay at the origin of the first or second pair of intercostal arteries beyond the coarctation, but occasionally were seen more peripherally. They were very thin-walled and easily damaged. Their origin from the aorta was often unusually large, in some cases measuring 2-3 mm. in diameter. Histologically the walls of these dilated intercostals were often deficient in muscle and elastic tissue, and in places were composed solely of intima and adventitia. They were dealt with either by ligation or by suture at their point of origin from the aorta. The ductus arteriosus was patent in five cases (Nos. 1, 17, 26, 29, and 31), widely in two and admitting only a fine probe in the others; in all it entered just above the coarctation. The left subclavian artery entered the narrow segment in one patient; at its point of entry its lumen measured 4 mm. and the artery was narrow from aorta to its first branch, where it abruptly widened to normal size. (The pre-operative brachial blood pressure was 210/105 in the right arm and 170/115 in the left arm.) In one patient the left subclavian artery arose so close to the coarctation that the origin of the artery was included in the anastomosis (see diagram, page 380). Vegetations as previously described were found on the interior of the resected aorta in eight cases.

Post-operative Course

The immediate post-operative course was uneventful in most cases. The death of one patient six hours after operation from rupture of a suture has been mentioned. Post-operative haemorrhage of severe degree occurred in two other cases but stopped spontaneously after transfusion.

In the majority of cases the blood pressure in the arms was near pre-operative levels in the immediate post-operative period for about 24 hours; it showed marked fluctuations, but gradually fell during the subsequent week or so to a lower and more stable level. Hypertensive crises have not been encountered. Cases with aortic valve lesions did not behave differently from the others.

In five cases abdominal pain in the post-operative period was severe enough to cause concern. In two cases it began on the fifth and in two others on the eighth post-operative day, was severe and colicky, and referred to the mid-abdomen but without localizing signs. In all four the pain subsided after several days. In the fifth case the pain was accompanied by a massive ascitic effusion. Resolution rapidly followed the removal of 2 litres of clear fluid. Price Thomas (1955, personal communication) has had similar experiences, and the signs demanded a laparotomy in an early case; the only abnormality seen was a little free fluid, mesenteric adenitis, and marked engorgement of the mesenteric vessels. Bowel function had not been disturbed in any of our patients, nor has there been any evidence of intestinal haemorrhage. There has been no evidence to suggest that the condition is due to mesenteric embolism.

Chylous fluid continued to drain from the left chest in one patient for several days after operation. The thoracic duct bears a close anatomical relationship to the aorta, and was doubtless damaged in the dissection. Cardiac tamponade occurred about a week after operation in one case in which the pericardium had been opened and pressure readings recorded from the left ventricle. Repeated aspiration of the pericardium was necessary for a few days, but the condition then subsided.

There have been no late complications in this series, but it has been our practice to delay ambulation for three weeks after operation and to restrict activities generally during the first six months.

Results of Operation

Mortality.—Three patients died. Two of these have already been noted: Case 19 had a dissecting aneurysm of the aorta, and the operation was advised as a desperate resort. In Case 6 death occurred six hours after operation from haemorrhage from the aorta due to rupture of the suture line. The third death, that of a woman aged 30 (Case 5), occurred suddenly five weeks after operation while she was in a convalescent home. At necropsy it was found that the suture line had parted and haemorrhage had taken place through the aperture in the aorta. Histologically, the aorta showed a cellular infiltration which suggested a low-grade infective process. These last-mentioned cases were the fifth and sixth of our series. It is well known that the mortality rates in any one clinic decrease with experience, and a mortality rate of less than 10% may now be anticipated in practised hands.

Follow-up.—The remaining 37 cases have been followed up after operation for periods ranging from three months to eight years, and all have remained in good physical health. Those patients who had symptoms from organic complications before operation have had no recurrences. Symptoms of cardiac anxiety states were alleviated in most of the cases in which they had been present pre-operatively, but were unchanged in three cases and worse in two. Both the latter had long-established neuroses, and one was a long-standing case of hysteria. Those patients who had syncopal attacks before operation had no recurrences after, but one patient developed occasional episodes for the first time post-operatively. No organic cause for them could be found.

Physical Findings During Follow-up.—The arterial pulsation in the neck always diminished. The femoral pulse increased in volume to within normal limits in all cases. The peak of the femoral pulse wave was reached either before or synchronously with the peak of the radial pulse in all but two cases. These findings were checked by arterial pulse records in 10 cases. Collateral arteries were no longer palpable. The murmurs were altered in all patients. The systolic murmur over the upper dorsal spine became fainter, but in only one case did it disappear completely. Its genesis and the significance of its persistence have been discussed above; it continued to be heard in cases that gave eminently satisfactory results on all other counts, and it could not therefore be used to assess the adequacy of aortic reconstruction. Otherwise the alterations in murmurs were as would be expected: the precordial systolic murmurs became faint, the early diastolic murmurs of aortic incompetence generally did not alter, and the mid-diastolic murmurs were less frequent. Only the apparent evolution *de novo* of aortic diastolic murmurs after operation in two cases was noteworthy.

Blood Pressure

The brachial blood pressure was reduced by the operation. The change was studied in detail in the first 20 patients in our hospital series, who were followed for three months or more after operation, and has been separately reported (Counihan, 1956). Both pre-operatively and post-operatively the brachial blood pressure fluctuated considerably, but the average of the post-operative pressures was lower by 32 mm. systolic and 16 mm. diastolic than the average of the pre-operative ones for the group. Statistically this fall is highly significant, the probability of its occurrence by chance being less than 1 in 1,000. However, the post-operative average brachial pressures were higher than the expected average pressures for the age and sex—that is, the pressures remained higher than normal.

No relationship was found directly between the amount of fall of pressure and the diameter of the aorta measured at the site of anastomosis. However, the amount by which the post-operative brachial pressure exceeded the expected average for the normal individual was apparently related

to the degree of deviation from uniformity in calibre of the reconstructed aorta, so far as systolic pressure was concerned, but not diastolic. The greater the restriction of the aortic diameter the more abnormal the systolic pressure. But even when uniformity was attained in the diameters of the reconstructed aorta the blood pressure remained higher than the expected normal.

The evidence that the residual hypertension was not due to the incomplete restoration of the aortic lumen in the cases studied is, first, all collateral vessels disappeared; secondly, the delay in the femoral pulse peak was abolished; and, finally, the femoral systolic blood pressure was at least as high as the brachial. The measurement of the femoral blood pressure by auscultation may be open to some inaccuracy, but from our figures after operation the femoral pressure, like the brachial, was above normal.

Many authors have found a generalized increase in the peripheral vascular resistance in coarctation. Furthermore, the fact that post-operative brachial pressures remained above the expected average for age is in harmony with the persistence of long-established hypertension after removal of a phaeochromocytoma or a diseased kidney. When a hypertensive agent has been acting for a sufficiently long time it appears that the blood pressure may become permanently set at high levels. We noted a tendency to a greater fall in the younger subjects than in the older; and, of course, the percentage fall has been greater in young subjects because their initial pressures were lower.

The three patients who had cerebrovascular accidents have had no recurrence and are in excellent health after a post-operative follow-up of 6½, 4½, and 2 years. The average post-operative blood pressure was in each case lower than the average pressure before operation. The reductions in average pressures were 54 mm. systolic and 5 mm. diastolic in one case, 20 systolic and 25 diastolic in another, and 58 systolic and 29 diastolic in the third, leaving these subjects with average pressures close to but somewhat above the expected average for their respective ages. In the one patient in whom papilloedema had been present it regressed after resection.

The Cardiogram.—When the cardiogram had shown evidence of left ventricular hypertrophy it was exceptional for it to return to normal; it did so in only three cases and showed a change towards normal in one other.

X-ray Appearances

The size of the left ventricle was unaltered in almost all cases. It appeared smaller in four but larger in one. The aortic arch retained a double contour in four, but was smaller in one of these. Aortic dilatation regressed except in one case in which the arch remained prominent. Where noted before operation, posterior oesophageal displacement disappeared except in two cases in which it remained substantially unchanged.

Rib-notching usually remained unchanged, although in one youth (Case 22) there has been marked regression.

Angiocardiography has been carried out post-operatively in only two cases, 9 and 20 months respectively after operation. One patient (Case 13) had an eroding post-stenotic aneurysm excised and grafted. The pre- and post-operative angiograms are shown in Figs. 5 and 6. In both of them the appearances of the aorta were satisfactory. The angiocardiograms of these two patients indicate that the reconstructed aortic lumen remains fully patent.

Discussion

Full assessment of the place of resection awaits a longer follow-up than is possible at present, but the encouraging results, so far, make a discussion of policy fruitful.

It has been shown that when organic symptoms occur in a person with coarctation serious complications have probably already developed. These may be remote from the coarctation (such as subarachnoid haemorrhage) or they

may occur locally (post-stenotic or intercostal aneurysms) and clearly indicate resection.

More often, however, the coarctation is accidentally discovered in a symptomless person, in whom the indications for surgery are less clearly defined. Investigations may reveal that trouble is afoot and that resection is desirable. But the finding of unsuspected vegetations and of thin-walled intercostal aneurysms at operation forbids complacency when investigations have been negative. Watching and waiting may end in disaster, with the occurrence of a vascular catastrophe. Alternatively, anxiety states are prone to develop during the waiting period. We believe that it is advisable to come to a decision without undue delay.

The menace from lesions in the neighbourhood of the coarctation has been successfully dealt with by resection in our cases. The occurrence of some of the more distant complications has been shown in this paper to depend, at least in part, on the hypertension. The reduction of this by resection has been demonstrated by detailed analysis of our first consecutive 20 cases that have been followed up for three months or more after operation. Our cases suggest that closest approximation to normal pressures is obtained in those patients in whom a uniform aortic lumen has been achieved, even though these pressures still remain above normal levels. It must be the surgical aim, therefore, to reconstruct the aorta with the maximum possible diameter. Marked residual constriction should be avoided by the use of grafts. The demonstrated fall in pressure does not seem to be large, but it should be noted that the figures refer to the average values of many pressure measurements taken at rest. They both reflect a constant lowering of the pressures and reveal the possibility that greater reductions have occurred in the blood pressure during daily activity as compared with levels obtaining before operation, since the blood pressure is said to rise excessively on effort in unresected coarctation (Gross, 1953).

Our three patients who had suffered from intracranial vascular accidents before resection had no recurrences afterwards. In the one in whom hypertensive neuroretinopathy had appeared, it regressed after operation.

Conclusion

The mortality of the operation is now small and the results have been satisfactory. The brachial blood pressure has been lowered towards normal. Post-operative cerebrovascular accidents, bacterial infection, aortic aneurysm, and heart failure have neither occurred nor recurred.

These are reasons for confidence that the bad prognosis for life carried by coarctations has been improved by operation, though this will be proved only by long follow-up.

The pros and cons of operation may be summarized as follows:—In favour: (1) the improbability of surviving past middle age; (2) the insidious or sudden nature of serious or fatal complications, such as aneurysms, endoaortitis, and cerebrovascular accident; (3) the reduction of blood pressure after operation; and (4) the low operative mortality. Against: (1) the difficulty of judging the risks to life in any individual patient; (2) the uncertainty that resection will prevent the onset of serious complications; and (3) the reluctance to submit a symptomless individual to a major operation of a prophylactic nature.

Serious consideration of this difficult problem has led us to believe that the pros outweigh the cons. Consequently we have recommended resection in all our cases below the age of 40 unless contraindicated by the presence of severe associated cardiovascular anomalies unlikely to be affected by the resection.

The ideal time for operation is before adolescence, because then the vessels are healthier and local vascular complications are less common. In addition, the likelihood of obtaining normal post-operative blood pressures is probably greater in the younger individual. Also, thoracotomy is

tolerated better by the young and anxiety states are unusual at this age. We believe, too, that a relatively serious operation of election should be carried out before the individual has acquired responsibilities and dependants.

Summary

Fifty-two cases of aortic coarctation have been studied.

A review of the literature and of our 12 unoperated cases emphasizes the poor prognosis in the untreated case.

Forty patients were submitted to operation.

Symptoms and physical and radiological signs have been evaluated before and after operation.

A surprisingly high incidence of unexpected local complications was encountered at operation, owing possibly to the preponderance of adults.

Three deaths occurred in the surgical series.

Satisfactory clinical results were obtained in the surviving patients, who have been followed up for from three months to eight years.

The brachial blood pressures have been reduced in all cases.

The factors influencing the post-operative blood-pressure levels have been discussed.

The indications for operation have been evaluated.

We are grateful to the many physicians who have so kindly referred cases to us, and especially to Dr. Paul Wood and Dr. Terence East for allowing us to quote patients who have been under their care before and after operation; to Dr. Wallace Bridgen, who most kindly allowed us to reproduce the pre-operative angiogram of Case 13; and to Professor J. McMichael, who gave valuable advice and criticism. We are also indebted to Mr. L. L. Bromley for permission to quote his operative findings in three patients. Finally, we must record our indebtedness to Sisters Anderson, Bowtle, and Prior for their invaluable and unstinting work in the wards and theatre.

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SURGICAL TREATMENT OF CHILDREN WITH COARCTATION OF THE AORTA

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Each advance in cardiovascular surgery increases the need for careful selection of patients suitable for operation. It is particularly difficult to come to a correct decision about cases of aortic coarctation because the disorder is compatible with a long and active life. Moreover, operation is not without risk, it does not always relieve hypertension, and even when the result seems at first to be successful an aneurysm may form at the line of anastomosis.

The difficulty is aggravated because when coarctation is first diagnosed it is usually impossible to predict the course of the disease, and, although undoubtedly some patients may be destined to attain a ripe old age, others will die before their due time. Sometimes death is sudden and without warning when the aorta or some smaller artery, such as one of those at the base of the brain, bursts under the strain; at other times death comes slowly when the heart fails or superimposed subacute bacterial infection proves resistant to treatment.

It might be possible to get a better idea of the likely outcome for the individual if the natural course of the untreated condition could be studied in a sufficiently large group of patients. But this again presents a problem, for during life the disorder must sometimes elude discovery, and after death, unless this occurs in hospital or is reported to the coroner, the body is unlikely to be examined by a pathologist. Probably many people with aortic coarctation die at home and are certified as having had hypertensive heart disease, myocardial infarction, or apoplexy. We recognize that these factors must make figures inaccurate and may lead to false conclusions. Nevertheless the selection of patients for operation will depend on criteria determined by experience, and we therefore record our experience of coarctation at the Birmingham Children's Hospital between July, 1950, and July, 1955.

Mortality

During these five years 30 infants (16 girls, 14 boys) died more or less directly as a result of coarctation, 17 within a week of birth, 8 within a month, and 5 within six months. Cases grossly complicated by associated defects have been excluded from this series. There was no record of a death from coarctation in an older child up to the age of 14, but we know of two deaths amongst adolescents with the disease. In this same period we saw at the hospital 22 children (14 boys, 8 girls) with coarctation, 21 of whom still survive.

It is impossible not to be impressed by the proportion of deaths occurring in infancy, especially in the first month. Fig. 1 (left-hand open column and solid columns marked A) shows that more than a third of Blackford's (1928) series of 323 cases of coarctation died in infancy. Most of these early deaths are associated with the infantile type of coarctation, but this is by no means invariable, and in our series one of the infants who died and two of the babies who have been operated on had the adult type of disease. Further, the infantile type of coarctation does not necessarily cause early death: four of the older children had a long narrow segment of aorta proximal to the origin of the ductus