COARCTATION OF THE AORTA: II. CLINICAL FEATURES

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Received January 15, 1947

This paper is concerned with the clinical features of coarctation of the aorta, and is based on a group of 26 cases observed by the author and described in the St. Cyres' Lecture, delivered on June 6, 1945. A paper on the collateral circulation in coarctation of the aorta has been published (Bramwell and Jones, 1941), and a further paper on the radiological diagnosis of the condition will follow: all three are based on the same series of cases.

Coarctation of the aorta has long been familar to anatomists, but until comparatively recently it generally eluded the clinician. In 1928 Maude Abbott was able to collect from the literature 200 cases over three years of age that had come to autopsy; but, of these, only 21 had been diagnosed during life as coarctation and 7 others as obstruction of the aorta. The same year Roesler published his classical paper on the radiological diagnosis of coarctation, and the following year Railsback and Dock published similar observations. Fray added a further paper in 1930.

These radiological observations provided the Mulberry Harbour that enabled clinicians to invade a territory hitherto dominated by morbid anatomists. Many cases have since been reported and I myself have seen 26* in the last fifteen years, 11 in private practice, 8 at my outpatient clinic at the Manchester Royal Infirmary, and 7 in recruits referred to me by medical boards of the Ministry of Labour and National Service during the war.

Autopsy statistics suggest that the incidence of this condition is about one per thousand. Fawcett (1905) found 18 cases in 22,316 autopsies at Guy's Hospital, an incidence of 0·08 per cent, and Evans (1933) 26 cases in 19,217 autopsies at the London Hospital, an incidence of 0·14 per cent; but I am inclined to think that the condition may be more common than these figures suggest, for I have reason to believe that coarctation sometimes escapes detection even at autopsy.

CLASSIFICATION

Most authors have followed Bonnet's (1903) classification and have described two types, the " infantile " and the " adult." In the former, which may be regarded as a persistence or exaggeration of the anatomical relations that exist before birth, there is a diffuse narrowing of the aortic isthmus. In the latter the narrowing, which is in the immediate vicinity of the insertion of the ductus arteriosus, may be so abrupt as to suggest that the aorta had been constricted by a ligature. The adult type was attributed by Craigie (1841) and some subsequent workers to an extension of the normal process that leads to occlusion of the ductus arteriosus after birth. This hypothesis is attractive, but there is considerable evidence in support of the view that the adult form also is due to an error in development at the junction of the primitive fourth and sixth branchial arches, an error that Maude Abbott suggests does not produce its effect till after birth when kinking of the descending limb of the aortic arch results from traction by the obliterated ductus.

* Five more cases seen since May 1945 will be reported later.

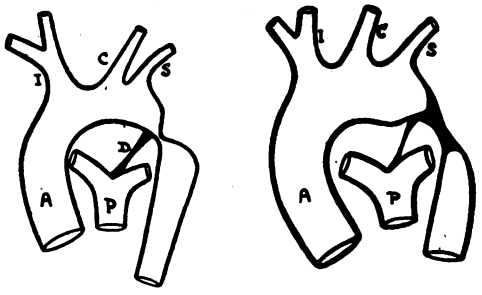

FIG. 1.—Coarctation: Evans's Types 2 and 3.

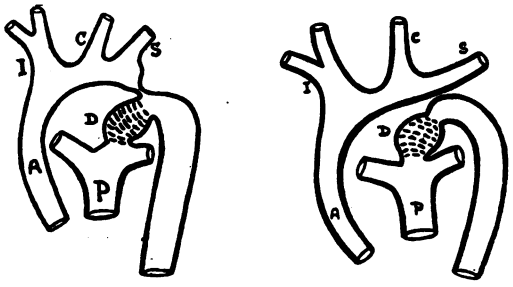

FIG. 2.—Coarctation: Evans's Types 1 and 4.

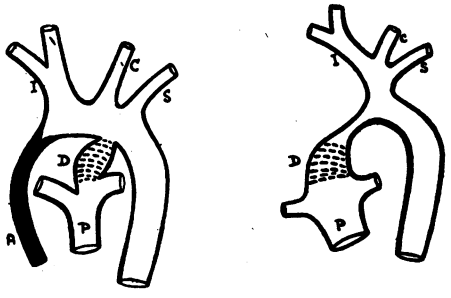

FIG. 3.—Coarctation: Evans's Types 5 and 6.

This classification, though useful, is not entirely satisfactory, and William Evans (1933) suggested an alternative classification based on purely topographical considerations. He described six anatomical varieties of coarctation of the aorta. I would suggest that Evans's classification might, without detriment, be simplified by dividing his six types into three groups. In Group I (Fig. 1), I would place Evans's Types 2 and 3 in both of which the ductus is closed and the proximal portion of the aorta is hyperplastic. Type 3 differs from Type 2 in that the coarctation is complete with atresia of the distal portion of the aortic arch, whereas in Type 2 there is merely a stenosis: this is a difference of degree rather than of kind. Similarly Group II (Fig. 2) would include Evans's Types 1 and 4, in both of which the ductus is patent and the proximal portion of the aorta is hypoplastic. These two types are essentially similar but differ from one another in that in Type 4 the interruption between the arch and descending aorta is complete, whereas in Type 1 there is a narrow communicating channel between them; again a difference of degree. Hypoplasia is rare compared with hyperplasia of the aorta and was present in only 10 per cent of Maude Abbott's cases. Lastly I would place Evans's Types 5 and 6 together in Group III (Fig. 3), since in both the only communicating channel between heart and aorta is the patent ductus. Admittedly there is an important developmental difference between these two types : in Type 5 the proximal portion of the arch is merely atresic or occluded, whereas in Type 6 it is absent.

The expectation of life (Table I) in these three groups is very different. In the first, in which the ductus is closed and the proximal aorta hyperplastic, 7 of Evans's 11 cases reached maturity; of 11 cases in the second group, in which the ductus is patent and the proximal aorta hypoplastic, 1 died in childhood and only 1 reached adult life; while in the third group none of the 6 cases survived infancy. The relatively favourable prognosis in Group I as compared with Groups II and III is probably, in part at any rate, attributable to the fact that the " infantile " type of coarctation is often associated with other grave errors of development, whereas in the so-called " adult " type of coarctation, in which there is an extreme degree of

TABLE I

PROGNOSIS OF EVANS'S SERIES

Group	Type (Evans)	Ductus	Proximal aorta	Coarctation	Age at death			Total
					Infant	Child	Adult	
I	2	Occluded	Hyperplastic	Partial	1	3	7	11
	3	Occluded	Hyperplastic	Complete				
II	1	Patent	Hypoplastic	Partial	9	1	1	11
	4	Patent	Hypoplastic	Complete				
III	5	Patent	Occluded		6	0	0	6
	6	Patent	Absent					
					16	4	8	28

stenosis, this is not the case. Maude Abbott found that of 82 cases in which death occurred within the first year, in 50 (61 per cent) the coarctation was associated with some other major congenital anomaly such as bilocular or trilocular heart, transposition of the arterial trunks, or pulmonary atresia, whereas in 155 cases in which life was prolonged beyond one year an associated major anomaly was present in only 13 (8 per cent) and, in these 13, coarctation was either so slight as to be of no importance or of the infantile type. In the older group, however, other minor congenital defects such as biscupid aortic valve, subaortic stenosis, or abnormal origin of the vessels arising from the arch of the aorta were common and occurred in 57 of the 155 cases.

Clinically, we may divide cases of coarctation into two groups in accordance with whether the ductus is closed or patent. In patients who survive infancy the former is much the more common. In Abbott's series of 200 cases the ductus was patent in only 18 and in my 26 in 3 or possibly 4 cases. The state of the ductus is important; since a patent ductus provides for the passage of blood from heart to aorta beyond the coarctation and thereby eliminates the necessity for a collateral circulation. Consequently, in such cases, there may be no pulsating vessels on the back or front of the chest and no notching of the ribs. This renders diagnosis more difficult. When the ductus is patent the upper part of the body is supplied by the branches of the aortic arch arising above the coarctation, while the trunk and lower limbs are supplied chiefly by blood entering the descending aorta through the ductus.

CLINICAL MATERIAL

Age. In Table II, I have arranged my cases in accordance with the age at which they first came under observation and have indicated their condition in May 1945: 13 were then alive, 10 had died, and 3 had been lost sight of. These figures are summarized in Table III.

Three of my patients were children, 3 were over 40 years of age, and the remaining 20 were between the ages of 18 and 37 at the time they were first seen. The small number of children in my series is due to the fact that both in hospital and in private practice the great majority of my patients are adults.

Sex. In Abbott's series of 200 cases, one-quarter were females and three-quarters males. If we exclude the 9 "Service cases," drawn from a special section of the population, almost entirely male, the ratio of the sexes in the remainder of my series is 6:11. Why the condition should be so much more common in the male sex it is difficult to say unless it be that, in reality, the sex incidence is equal; but, since women lead a more sheltered life, they are less exposed to the physical strains so often responsible for the development of heart failure in males.

TABLE II

BRAMWELL'S SERIES

Case No.	Sex	Age first seen	After history 1945	
			Age living	Age at death
1	M	9	—	11
2	M	11	—	23
18	M	11	—	12
16	M	18	21	—
20*	M	18	20*	—
22	M	18	19	—
25	M	19	19	—
15	M	20	—	23
14	M	21	25	—
11	M	23	lost sight of	—
24	F	23	—	23
13*	M	24	29*	—
8	F	25	—	27
21	M	26	lost sight of	—
12	M	27	lost sight of	—
23	M	27	28	—
17	M	30	38	—
7	M	31	45	—
4	M	31	39	—
9	F	31	—	36
10	F	32	39	—
19	M	32	35	—
26	F	37	37	—
5	M	46	—	52
3	F	51	—	52
6	M	58	—	62

* See p. 125.

TABLE III

AGE AND PROGNOSIS OF BRAMWELL'S SERIES

Ages in 1945 or at death	Living	Dead	Untraced	Total
10–19	2	2	—	4
20–29	5	4	3	12
30–39	5	1	—	6
40–49	1	—	—	1
50–59	—	2	—	2
60–69	—	1	—	1
Totals	13	10	3	26

This would render it more likely for the condition to remain latent in the female and to pass unnoticed unless it were discovered accidentally in the course of some routine examination. This hypothesis, however, would not account for the high incidence of males in Abbott's series in which the figures are based on autopsy statistics, but for the fact that autopsies are performed less often in the female than in the male. Dr. W. Susman (1946) informs me that in a consecutive series of 2000 autopsies performed at the Manchester Royal Infirmary 63 per cent were males and only 37 per cent females.

Service Cases. Nine of the young adults must be placed in rather a different category from the others since they would not have come under observation but for the war. The lesion in these men was discovered in the course of a routine examination and most of them had been unaware of any disability and had no cause to seek medical advice. Seven were recruits referred to me by recruiting medical boards, one (Case 25) consulted me privately having been rejected for service, and one (Case 21) was a serving soldier referred to my clinic.

TABLE IV

DETAILS OF "SERVICE CASES"

Case	Age	Symptoms	Athletic	Remarks
14	21	Dysp. 2–3 years	+ till 19	
11	23	—	+ at school	
15	20	Septicæmia	+	Aortic incompetence
16	18	Dysp. and pain 2 years	+ at school	
19	32	—	Heavy work	Dyer
20	18	—	++	Swim, rugger, cycle, climb
21	26	Epistaxis	—	Serving soldier
22	18	—	Heavy work	Patent ductus
25	19	—	+	

Six of these men were free from symptoms and had been unaware of any disability prior to the discovery of their cardiac lesion at the medical board. Of the other three, one (Case 14), aged 21, had played football up to the age of 19, but subsequently complained of tiredness and some breathlessness on exertion: another (Case 16), a lad of 18, had complained of dyspnœa and pain in the calves for two years but had played games at school. The third (Case 21) had been known to have heart trouble from childhood, and had always avoided strenuous exertion since it was liable to cause epistaxis.

TABLE V

PERLMAN'S SERIES OF AMERICAN ARMY RECRUITS

Case	Age	Heart disease diagnosed	Athletic
1	28	—	+
2	21	—	+
3	21	—	?
4	19	—	?
5	22	14	++
6	21	early childhood	0
7	20	—	+
8	20	11	0
9	18	—	++
10	23	8	+
11	32	22	+
12	25	early childhood	0
13	18	—	+

It is interesting to compare this group with the series reported by Perlman (1944) from American Army recruits. The ages of the two groups are very similar, 10 of Perlman's cases and 7 of mine being under 25 and none over 33. Seven of Perlman's cases and 6 of mine had no knowledge of heart disease and were free from symptoms; 8 in each series had participated actively in athletics or had been employed in laborious physical work. One of my patients (Case 20) was a good athlete and had gone in for swimming and played Rugby football at the

Manchester Grammar School: he also was a keen cyclist and climbed in Switzerland. One in Perlman's series played semi-professional basket-ball.

Symptoms

The age at which patients first come under observation is a matter of chance, since some seek medical advice for symptoms attributable to their coarctation, others for symptoms due to some intercurrent disease, and in others, unaware of any disability, the lesion is discovered accidentally in the course of a routine examination. The age at which symptoms first appear however, does seem to be of significance in prognosis and I have accordingly classified my cases in three groups.

1. In 5 symptoms dated from childhood. Of these, 2 died in the second decade, 2 in the third, and 1 has been lost sight of.

2. In 11 patients symptoms first appeared between the ages of 16 and 57. Four of these are still alive aged 21, 25, 29, and 37 respectively; 1 has been lost sight of; and the other 6 are dead.

3. Of the remaining 10 patients 1 has been lost sight of and, up to May 1945, the other 9 were either free from symptoms or complained only of symptoms due to some disease unconnected with their coarctation. Their age incidence was from 19 to 45.

Cases in which Symptoms Dated from Childhood

Case 1. A boy, aged 11, was referred to me in December 1934 by Dr. John Ward from the Royal Manchester Children's Hospital to which he had been admitted with a sore throat. Prior to severe epistaxis at the age of 9 he had been free from symptoms. In addition to coarctation, he had subaortic stenosis with great cardiac enlargement (see p. 112). He was readmitted to hospital in May 1936 with acute pericarditis with effusion following a further attack of tonsillitis. From this he made a good recovery, but a few months later developed congestive heart failure from which he died.

Case 8. A woman of 25 consulted me regarding her fitness for pregnancy. Her chief complaint was pain in the back. Two years later she developed subarachnoid hæmorrhage from which she died (see p. 106).

Case 18. This boy appeared to be not only healthy but robust until, at the age of 11, he developed subacute infective endocarditis from which he died. There was evidence, both clinical and radiological, of a patent ductus (see p. 115).

Case 21. A serving soldier was referred to my clinic by Major Olav Kerr, R.A.M.C. Since childhood he had always avoided strenuous exertion since it was liable to produce epistaxis. After three years' service as a draughtsman he decided to take a sapper's course for promotion, but this proved too strenuous for him and provoked dyspnœa. Shortly afterwards he was invalided out of the service following pneumonia and has since been lost sight of.

Case 24 (see p. 113). A woman of 23 had been short of breath as long as she could remember and cyanosed in cold weather. While in hospital she had two peculiar attacks in which she complained of paræsthesia in the lower limbs associated with a feeling of constriction in the lower sternal region. Following her second attack she developed œdema and died of congestive heart failure. In this case the presence of a widely patent ductus (Fig. 6) was confirmed post-mortem and the coarctation was of the infantile type.

Cases in which Symptoms Developed Later in Life

Case 2. A boy of 11 was referred to me in 1930 on account of palpitation following a minor gastro-intestinal disturbance and was found to have coarctation of the aorta. Throughout his time at the Manchester Grammar School he had no illness, and played cricket and swam. He remained free from symptoms till 1942 when he developed subacute infective endocarditis from which he died.

Case 3. A woman aged 51 first noticed tightness in the chest and palpitation at the age of 50. She had worked hard all her life and had never required medical attention previously, but had always preferred a standing job as she got pain in the left chest if she had to sit for long (see p. 107).

Case 5 (see p. 107). A man, aged 46, was referred to me complaining of a pain in the right arm of twelve months' duration. When seen again four years later the pain had ceased to trouble him and he died of carcinoma of the stomach at the age of 52.

Case 6 (see p. 112). In March 1933, I was consulted by a man of 58 who had enjoyed good health and led an active life until twelve months previously, when he began to suffer from dyspnœa on exertion with a sensation of constriction in the chest. Six months later he developed auricular fibrillation and shortly afterwards had a severe coronary occlusion. In June 1935 he was still fibrillating but free from symptoms and able to carry on his business. He died of lobar pneumonia at the age of 62. This man's freedom from symptoms was remarkable as, in addition to his coarctation, he had aortic incompetence and his heart was greatly enlarged. Two years before death he underwent a major operation from which he made an excellent recovery.

Case 9 (see p. 122). A woman, aged 31, first noticed shortness of breath in the fourth month of pregnancy. She was successfully delivered by Cæsarean section. Two years later she again became pregnant and developed auricular fibrillation. She died from congestive heart failure at the age of 36.

Case 12. A varnish maker, aged 27, was referred to me by my colleague, Dr. A. Hillyard Holmes, whom he consulted on account of tiredness and attacks of vomiting a month previously. Only since this attack had he been short of breath and suffered from palpitation. This case has been lost sight of (see p. 107).

Case 13. A storekeeper, aged 24, was referred to me on account of high blood pressure. He had suffered from epistaxis since the age of 13, but had played football regularly until a year before I saw him, in spite of the fact that since 17 he had complained of pain in the back which troubled him intermittently. As this man's only symptom prior to the age of 17 was epistaxis, and since he had participated actively in athletics up to the age of 23, I have not included him in the group of cases in which symptoms dated from childhood (see p. 107 and 125).

Case 14. A leather worker, aged 21, who had been short of breath and easily tired for 2 or 3 years, prior to which he had been perfectly well and played football, was referred to me by a recruiting medical board in 1940. In May 1945 he was feeling fit and still anxious to join the Navy. In this case it is very doubtful whether the temporary asthenia in 1938–40 was attributable to his coarctation.

Case 15. A shop assistant, aged 22, was referred to me by a medical board in April 1941. He had always enjoyed excellent health and gone in for football, cricket, rowing, and cycling. He was admitted to Crumpsall Hospital in March 1944 and I am indebted to the Superintendent, Dr. Ramsay, and to Dr. R. W. Luxton, for particulars of his illness at that time. Blood culture on two occasions yielded *Staphylococcus aureus* and he was considered to be suffering from septicæmia. He died five days later, but unfortunately permission for a necropsy was not obtained. This young man never had any symptoms until ten days before his death. His coarctation was unsuspected at the time of his acute fatal illness, which may have been acute infective endocarditis.

Case 16. An accountant's clerk, aged 18, who had played tennis and cricket at school, began to complain of breathlessness and pain in the calves on exertion at the age of 17. He was referred to me by a medical board in January 1942: in May 1945 he informed me that though he had occasionally complained of tiredness he had not been off work.

Case 26. A woman, aged 37, complained of slight dyspnœa on exertion, but otherwise, apart from an illness 17 years previously, had been free from symptoms excepting during her two pregnancies (see p. 121).

Discussion of Symptoms

To summarize these findings, the symptoms of which my patients chiefly complained were as follows:

Three (Cases 2, 15, and 18) were free from symptoms until they developed infective endocarditis or septicæmia, three (Cases 1, 13, and 21) complained of repeated epistaxis, and one (Case 8) of migraine dating from childhood; it is interesting to note that she died of subarachnoid hæmorrhage. Breathlessness and palpitation on exertion were common symptoms as was undue tiredness. Eight of my patients complained of pain which affected various parts of the body. This symptom merits more detailed consideration.

Case 8. A parson's wife, aged 25, consulted me in July 1938 regarding her fitness for pregnancy. In childhood she suffered from migraine and at school she was never able to play games because of

dyspnœa and palpitation. From 17 until her marriage at 24 she worked in an office and was never absent owing to illness except for occasional sore throats. When I saw her she was able to do all her housework with the help of a woman on one half-day a week, but complained of pain below the angle of the left scapula aggravated by exertion and occasional dyspnœa and palpitation. Apart from the pain in the back, she remained fairly well until 1940, when she had a subarachnoid hæmorrhage from which she died. The post-mortem findings in this case have been fully reported (Bramwell and Jones, 1941).

Abbott (1928) has called attention to the high incidence of subarachnoid hæmorrhage from rupture of a congenital cerebral aneurysm in patients with coarctation of the aorta.

Pain was the presenting symptom in four other cases in this series.

Case 3. The oldest woman in my series, first seen at the age of 51, had worked hard all her life and had been free from symptoms until twelve months previously; but since childhood if she sat for long, especially when leaning forward sewing, she got a pain in the left sub-mammary region, and for this reason she said she had always chosen a " standing-up job." It is difficult to explain the association of the pain with the sitting posture in this case. From the age of 14 to 35 she worked as a weaver and after that as a maker-up in a warehouse. This employment involved lifting heavy packages. Three years before I saw her, the warehouse closed down, and since then she had been doing housework and taking a child to school each day. She had never required medical attention except for colds until she developed lichen planus at the age of 50. She then began to complain of tightness in the chest and pain similar to that which she had experienced when sitting for long. At that time she had venous engorgement in the neck but no œdema. A few months later she developed congestive heart failure from which she died.

Case 5. A medical practitioner, aged 46, was referred to me in January 1934 by my colleague, Dr. Fergus R. Ferguson, whom he had consulted the previous November for an aching pain of twelve months' duration, felt in the region of the insertion of the right deltoid, the outer side of the forearm, midway between the wrist and elbow, and in the tips of the fingers. It occurred at any time of the day or night, but was aggravated by using the arm driving his car which had a right-hand gear change. For twelve years he had been subject to cramp in the right arm and for five years his handwriting had been somewhat shaky. Having had chorea at the age of 14, he had been stopped playing games at school on account of his heart, but later played tennis. Following influenza in 1923 he had suffered from asthma until 1930. I obtained no history of other illness. His biceps and supinator jerks were diminished on the right side, but the triceps jerk was normal. On a neurological basis, Dr. Ferguson considered that the signs and symptoms could only be accounted for by the somewhat unsatisfactory diagnosis of " brachial neuritis " and he was inclined to think that pressure from the dilated arteries on the nerves of the brachial plexus was a more plausible explanation. This, however, seems doubtful, for when seen again in May 1938 he stated that the pain in the arm had ceased to trouble him. He eventually died of carcinoma of the stomach at the age of 52.

Case 12. A man of 27 complained of attacks of præcordial pain passing through to the back, on account of which he was suspected of aneurysm.

Case 13. A storekeeper, aged 24, began to complain of pain in the back at the age of 17. Nevertheless he continued to play football regularly until twelve months before I saw him. He then gave it up, not on account of the pain, but because it made him unduly tired. The pain was most severe when he first got up in the morning and passed off in about half an hour. At the age of 22 he had a course of treatment from an osteopath over a period of 18 months and was then told he had a high blood pressure. Hence he consulted me. When I last heard of him five years later he stated that he was feeling quite well. The introspective nature of this patient made me inclined to discount his symptoms to some extent.

In two of the four cases reported by King (1926) pain was a prominent symptom. One of his patients, a man of 35, complained of pain on exertion in the left side of the chest, and a year later in the right chest. In King's other patient, a man of 58, the pain was in the left supra-clavicular fossa and left shoulder. One of Evans's (1933) patients complained of pain along the left border of the scapula. In both these cases the pain corresponded in position to areas of arterial pulsation.

Enlarged collateral channels might produce pain in several ways. Evans (1933) suggested

that pain in the back in patients with coarctation might be due to erosion of ribs by the enlarged intercostal arteries, like the pain produced by an aneurysm of the aorta which erodes the vertebræ, but the frequency with which well-developed rib notching is present without pain makes this explanation improbable.

Pain in the arm or around the costal margin is more suggestive of root or nerve pressure. This might be due either to direct pressure on nerve trunks or to pressure by the enlarged anastomotic artery where it passes through the intervertebral foramen.

The importance of the spinal anastomosis in coarctation of the aorta has not been generally appreciated. The vertebral artery, arising from the first part of the subclavian, reinforces the spinal arteries in which the blood flows downwards to reach the spinal branches of the aortic intercostals. These pass through the intervertebral foramina. There are also branches from the inferior thyroid artery which pass through the intervertebral foramina in the neck to join the spinal arteries.

A patient reported by Haberer (1903) is of unusual interest in this connection. Three days before admission to hospital she developed transverse myelitis from which she died three months later. At autopsy it was found that the anterior spinal artery had contributed to the collateral circulation and had caused compression of the spinal cord at the level of the second dorsal segment. In spite of atresia of the aorta, this woman had borne seven children.

The association of the pain with exertion can be explained by the increased blood flow through the tortuous and pulsating collateral channels, and the differing sites of pain may be related to differences in the anastomotic pattern.

In view of the low blood pressure in the lower limbs, one would have expected intermittent claudication to be common in patients with coarctation of the aorta, but only two of my patients complained of pain or paræsthesia in the legs—an army recruit who had played games at school, and had complained of pain in the calves on exertion since the age of 17 (Case 16) and a young woman with coarctation and a patent ductus who suffered from peculiar attacks of paræsthesia, associated with a sensation of heaviness but not actual pain in the lower limbs (Case 24).

In both cases reported by Blumgart, Lawrence, and Ernstene (1931) occasional cramp in the legs was complained of. King (1926), Parsons-Smith (1921), and Woltman and Sheldon (1927) each reported a case with intermittent claudication and in Gossage's case (1913) the patient suffered all his life from weakness in the legs.

Cases in which there were no Symptoms

It is surprising how many patients with coarctation of the aorta are not only free from symptoms, but reach a high standard of physical and mental development. Maude Abbott refers to the case of a university professor " who presented no signs of failing circulation unless his remarkable mental development and an unusually lively and restless nature were signs of an arterial hyperæmia of the brain."

Nine of my patients were free from symptoms in May 1945 and of these 5 were over 30 years of age. In addition there were 3 (Cases 6, 3, and 5) who reached the ages of 57, 50, and 45 respectively before symptoms developed. Three young recruits (Cases 20 (see p. 125), 22, and 25) and the following 6 older patients are known to have been symptom-free up to May 1945.

Case 4. A man, now 39 years of age, who has had no symptoms directly attributable to his coarctation, was referred to me in 1937 by my colleague, Professor E. D. Telford, whom he had consulted on account of incipient gangrene of the terminal phalanx of the middle finger of the left hand. This he attributed to an accident two years before in which the nail had been torn off: examination, however, revealed a predisposing local lesion in the left arm, namely congenital phlebarteriectasis. In 1938 he was admitted to the Middlesex Hospital where the affected finger was amputated. When I last saw him in May 1945 he stated that since 1941 he had held a clerical job and had been off work

only one day. The left arm still ached at times and this occasionally kept him awake at night. The aching was relieved by elevating the limb. The symptoms in this case may have been entirely attributable to his phlebarteriectasis.

Case 7. A business man, who is now 45, never had any symptoms apart from occasional extrasystoles. He first consulted me in 1931 as high blood pressure had been discovered in the course of a routine examination. There was nothing of significance in his medical history and he was able to play a hard game of tennis. When I last heard of him, in 1945, he stated that he had not felt better for years. He worked hard during the war, his hours were long, and he could dig for long periods in his garden—a large one which he kept entirely unaided.

Case 10. This woman is living and well at the age of 39 (1945). She was referred to me on account of thyrotoxicosis in 1938. Before this she had been very fit and during the summer of 1937 had played tournament tennis. The thyrotoxicosis was cured by a course of X-ray treatment. Since then she has been free from symptoms and during the war has engaged in Red Cross work in addition to her housework and has undertaken the active charge of a large garden which has entailed a lot of heavy digging (see p. 112).

Case 17. An assistant works manager in a big industrial firm, now aged 38, consulted me in 1937 having been refused a pilot's licence on account of high blood pressure, though he had previously served in the Oxford University air squadron. He was able to play tennis and squash without the least discomfort, and during the war he became a platoon sergeant in the Home Guard, but retired after three years' service on account of insomnia, which he wisely regarded as a warning to avoid violent exertion. Since then he has had no further trouble and has been able to deal with the strenuous conditions of war-time factory management.

Case 19. A dyer, aged 32, who was employed during the war as a foundry labourer, was referred to me by a medical board in 1942. All his life he had done laborious work and had never been conscious of any disability. He wrote me in 1945 saying that he still kept perfectly fit. He was working seven days a week and would have liked to get into the army.

Case 23. A man, aged 27, consulted me on account of a murmur, due to a patent ductus. He also had coarctation of the aorta, but neither lesion had ever caused any disability. (See p. 115.)

To sum up, 9 of the 23 cases which it has been possible to follow up were free from symptoms in May 1945 and 5 of these were then over 35 years of age. In addition 3 other patients in my series did not develop symptoms till after the age of 45.

To look at the problem from another angle, if one excludes the 3 who have been lost sight of, the remaining 23 in my series may be divided into three groups in accordance with their age at the present time. Three lived to over 50 years of age. All three are now dead, but two died of conditions unconnected with their coarctation, namely carcinoma of the stomach and lobar pneumonia. None of them had any symptoms before the age of 45 and one lived to 62. Between 30 and 50 there are 7 cases. Of these one died at 36, after her second confinement; the other 6 are alive and 5 have been free from symptoms up to the present time. Of the 13 cases under 30 years of age, 6 have died and 7 are living, 4 of the latter being free from symptoms.

TABLE VI

PROGNOSIS OF AGE GROUPS (BRAMWELL)

	Dead	Symptoms	No symptoms	Total
Over 50	3	—	—	3
30–50	1	1	5	7
10–30	6	3	4	13
Totals	10	4	9	23

The third decade appears to be the dangerous period, and this is not surprising for it is then that the men are exposed to the greatest physical strain and the women are likely to have their first pregnancy, while subacute infective endocarditis is common at this age.

The number of patients over 30 years of age who are not merely free from symptoms but able to undertake considerable physical activity suggests that when the coarctation is sufficiently well compensated to surmount the hazards of the third decade it may cause little trouble until the degenerative period of life.

Diagnosis

The classical signs of coarctation are (1) high arterial pressure in the upper limbs associated with lower pressure in the lower limbs, (2) pulsating arteries on the back or front of the chest, and (3) rib notching as shown by radiography. One or more of these signs may be absent: in some cases there is no clinical evidence of a collateral circulation and the ribs are not notched, and occasionally the blood pressure is not above normal.

To these signs I would add two others that are unlikely to escape detection in the course of an ordinary routine examination—excessive arterial pulsation at the root of the neck and a systolic murmur, the distribution of which does not conform to that of the common valvular or congenital lesions; this murmur may be very clearly heard, in fact it is sometimes loudest, in the interscapular region. There is also a radiographic abnormality that may be of diagnostic importance when rib notching is absent—a double aortic knuckle.

The Arterial Pressure. As Lewis (1934) pointed out, unless the possibility of coarctation be kept in mind in all cases of unexplained hypertension, the condition is apt to escape recognition, and palpation of the femoral pulse should be included in the routine examination of all such cases. I would add that in order to avoid missing coarctation the blood pressure in the lower limbs should be taken in all cases of patent ductus and in those cases of aortic incompetence in which the ætiology is obscure.

Occasionally the blood pressure in the upper limbs is not raised. In Case 10 in my series it was 140/90 and in Case 2 it fell from 165/90 to 130/60 when subacute infective endocarditis supervened. In one of the children in my series (Case 1) in whom the coarctation was associated with subaortic stenosis the pressure was only 95/75. Hallock and Hebbel (1939) reported a case without hypertension: in one of King's (1937) cases the blood pressure was only 123/87 and in Case 12 of Blackford's (1928) series, a man of 24 who had served in the American Navy, the pressure was 128/88.

The Collateral Circulation. The collateral circulation in coarctation of the aorta was first described by Paris in 1791. Unless the back and front of the chest be inspected carefully in a good light, pulsating arteries can easily be overlooked. In one of my patients (Case 9) in whom the collateral circulation had been quite obvious when the heart rhythm was normal, pulsation could only be detected by very careful palpation when auricular fibrillation supervened.

A late systolic murmur is generally audible over the dilated arteries that form the collateral circulation and may be heard on the back of the chest even when no pulsation is palpable. When the internal mammary and deep epigastric anastomosis is well developed, the peculiar distribution of the murmur, which is equally well heard on both sides of the chest in an area parallel to the border of the sternum, can hardly fail to attract attention.

The superficial anastomosis is no sure indication of the extent of the anastomosis as a whole. In Case 8 in our series, which came to autopsy, one of the most important anastomoses proved to be between the superior intercostal, arising proximal to, and the aortic intercostals, arising distal to the coarctation. This anastomosis is deeply situated and is inaccessible to palpation. It is not surprising that these vessels should be greatly dilated since they provide the shortest route to circumvent the coarctation, and when this route is freely patent the femoral pulse may be of good volume even though there be complete atresia of the aorta. When on the other hand the blood has to follow a more circuitous route to reach the descending aorta, the volume of the femoral pulse is correspondingly reduced; for, as Bonnet pointed out, the

sensation conveyed to the finger by palpation of the pulse depends, not on the blood flow, but on the steepness of the pressure gradient along the front of the pulse wave: the volume of the femoral pulse bears no relation to the degree of stenosis of the aorta.

In normal subjects the femoral and radial pulses are synchronous on palpation, but in many cases of coarctation the femoral pulse appears to be delayed. This is due to the fact that it is not the arrival of the pulse wave in the artery, but the crest of the wave that is appreciated on palpation and, in coarctation, the pulse in the lower limb is of the slowly rising type.

Other problems relating to the collateral circulation are more fully discussed in our previous paper (Bramwell and Jones, 1941).

Radiological Diagnosis

The discovery that rib notching could be demonstrated radiographically was a great stimulus to the diagnosis of coarctation of the aorta. Prior to the publication of Roesler's paper very few cases had been recognized during life. Rib notching does not occur in every case, and is absent when the coarctation is associated with a freely patent ductus arteriosus. Well-developed rib notching was present in 17 of the cases in my series: in 5 (Cases 11, 17, 20, 21, 22) it was not sufficiently definite to be of diagnostic value and in 4 it was absent; two of these (Cases 1 and 18) were children and in 3 of the 4 (Cases 18, 23, and 24) the ductus was patent.

Laubry's (1937) suggestion that rib notching is not pathognomonic of coarctation led us to make further observations on the point. These have convinced us that the trivial degree of notching occasionally met with in other conditions is easily distinguished from the notching of coarctation.

The absence of the aortic knuckle has been stressed by most writers as an important radiological sign, the vascular pedicle being funnel shaped; but, although this is a striking feature in some cases, in others, prominences that may easily be mistaken for the aortic knuckle are produced by other structures.

Direct demonstration of discontinuity of the aorta with the patient in the left oblique position is theoretically the most convincing proof of coarctation, but in our experience this is rarely possible even by the use of tomography. Fray (1930) states that a defect in the aorta can be demonstrated in all cases in which it is possible to obtain a satisfactory left oblique film, but his statement appears to be based on only two cases. Taylor (1934), on the other hand, was unable to demonstrate discontinuity in any of her five cases, Roesler (1943) agrees that it is sometimes impossible, especially in young subjects, and Brown (1939) states that even in the adult it can rarely be demonstrated.

When rib notching is absent, the most important radiological sign of coarctation is the presence of a double prominence in the region of the aortic knuckle (Fig. 4 and 7), the upper component being formed by the dilated left subclavian artery where it arises from the blind end of the aorta proximal to the coarctation, and the lower component by the blind end of the descending aorta. This and other aspects of the radiological diagnosis will be more fully discussed in a subsequent paper.

In five of the cases in the present series the diagnosis was made by the radiologist (Table VII). In one (Case 6) the clinical signs were attributed to associated aortic regurgitation, in another (Case 10) in which the blood pressure was only 140/90 to Graves's disease, while in a third (Case 1) they were overshadowed by those of subaortic stenosis. It is in cases such as these, in which some additional lesion is present, that coarctation is especially liable to be overlooked. The other two cases were referred to me by my colleagues after the condition has been diagnosed radiologically—Case 5, with pain in the arm which it was thought might

TABLE VII

CASES DIAGNOSED BY RADIOLOGY

Case	Sex	Age	Clinical diagnosis	Radiology
6	M	58	Aortic incomp.	Rib notching
10	F	31	Thyrotoxicosis	Rib notching
1	M	9	Subaortic stenosis	Double knuckle
5	M	46	? Cervical rib	Rib notching
12	M	27	? Aneurysm	Rib notching

be due to a cervical rib, and Case 12, where the physical signs suggested the possibility of aneurysm of the aorta.

In Case 6, aged 58, the heart was greatly enlarged, he had an aortic diastolic murmur, and the blood pressure was 200/80. No subcutaneous arterial anastomosis was detected, but X-ray examination showed notching of the ribs and absence of any aortic knuckle or aortic impression on the œsophagus. Clinically this appeared to be a straightforward case of aortic incompetence with auricular fibrillation, but there was one feature that should have led us to suspect coarctation—the high systolic pressure. In aortic incompetence due to rheumatic infection, syphilis, or arteriosclerosis, the systolic pressure rarely exceeds 170.

In Case 10, referred on account of thyrotoxicosis, arterial pulsation in the neck was excessive but this did not arouse my suspicion, as I attributed it to the overacting heart, and the blood pressure was only 140/90. I referred her to Dr. E. W. Twining for X-ray treatment and it was he who made the diagnosis. Thinking that the carotid pulsation was more than could be accounted for by the thyrotoxicosis, he took a film of the chest which showed typical rib notching. This was the only adult patient in my series in whom the blood pressure was not at any time found to be above normal.

The association of thyrotoxicosis with coarctation is interesting. Cookson (1936) who reported a case of coarctation of the aorta with toxic goitre, treated by complete thyroidectomy, suggested that the association is too frequent to be fortuitous. In support of this hypothesis, he refers to papers by Loriga (1887), Blackford (1928), Ulrich (1931), Amberg (1932), Eppinger and Midelfart (1933) and Brown (1934). Of the 9 cases reported by these authors, 7 were females. Cookson suggests that the mechanism by which thyroid disturbance arises in association with coarctation is the increased blood supply to the gland, as a result of the collateral circulation through the superior and inferior thyroid arteries which arise from the external carotid and subclavian respectively, above the coarctation.

In the fifth case, diagnosed by radiology (Case 1), the apical cardiac impulse was in the anterior axillary line and a second impulse and systolic thrill were palpable to the right of the manubrium. A to-and-fro murmur was audible over the manubrium and a systolic murmur on the back of the chest. The blood pressure was 95/75 and the femoral pulse was of good volume. Dr. Evan Bedford kindly gave me his opinion on the interpretation of the unusual X-ray appearances in this case (Fig. 4). He considered that the double aortic knuckle signified coarctation of the aorta, the upper knob corresponding to the blind end of the aortic arch and the lower one to the blind end of the descending aorta. My colleague, the late Dr. E. W. Twining made an exhaustive tomographic investigation of this case and satisfied himself that this explanation was correct. I have since been struck by the extraordinary similarity of this radiogram to that in Hamilton and Abbott's (1928) case.

COARCTATION ASSOCIATED WITH PATENT DUCTUS

The diagnosis of coarctation when associated with a patent ductus may be very difficult.

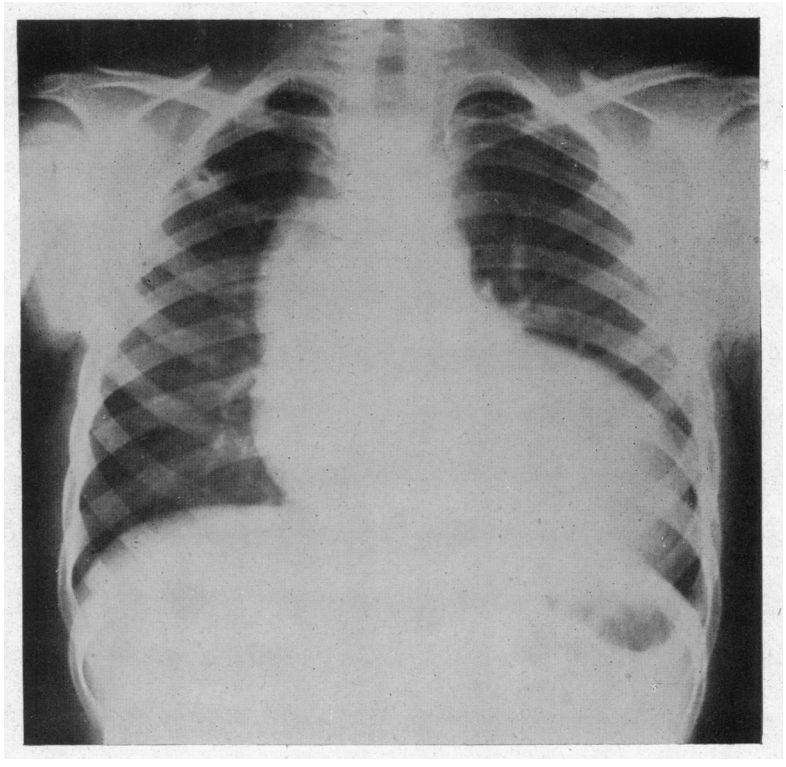

FIG. 4.—Radiogram Case 1, showing double aortic knuckle (see text).

The first case was diagnosed post-mortem, the second clinically, and in the third, a child, the diagnosis was suspected.

Case 24. An unmarried woman, aged 23, was admitted to hospital from my outpatient clinic on November 26, 1941, complaining of dyspnœa and œdema of the feet. She had been short of breath as long as she could remember and cyanosed in cold weather since birth. These symptoms had been worse during the past year. In July 1941 she had been admitted to Withington Hospital with retention of urine and acute respiratory distress and I am indebted to the Medical Superintendent, Dr. Greenwood, for information regarding her condition at that time. She then had œdema of the feet and complained that she had " no use in her legs." Her symptoms were relieved by catheterization.

The heart shadow (Fig. 5) was globular with generalized cardiac enlargement, but no rib notching. Auscultation revealed triple rhythm at the apex and a harsh murmur extending from systole into diastole, but chiefly diastolic, maximal in the third left intercostal space close to the sternum. Moist sounds were present at the bases of the lungs, and the liver was slightly enlarged. Her systolic pressure was 170, but the diastolic end-point was indefinite.

On November 28 she complained of tingling starting in the left foot, then affecting the right foot, spreading up to the thighs, and also involving the left arm. This symptom had occurred on previous occasions. It was associated with a feeling of constriction in the lower sternal region. The legs felt heavy, but were not actually painful, and during the attack the dyspnœa and cyanosis were accentuated. All pulses were palpable in the arms and legs, but that in the dorsalis pedis was very feeble. Catheterization again relieved her symptoms and on the following day her condition had returned to normal. On December 1, she had another similar attack with paræsthesia in the lower part of the body. This time catheterization and morphia gave only incomplete relief. Her condition progressively deteriorated and she died three days later.

The weight of the heart post-mortem was 600 g. Both right and left ventricles were greatly

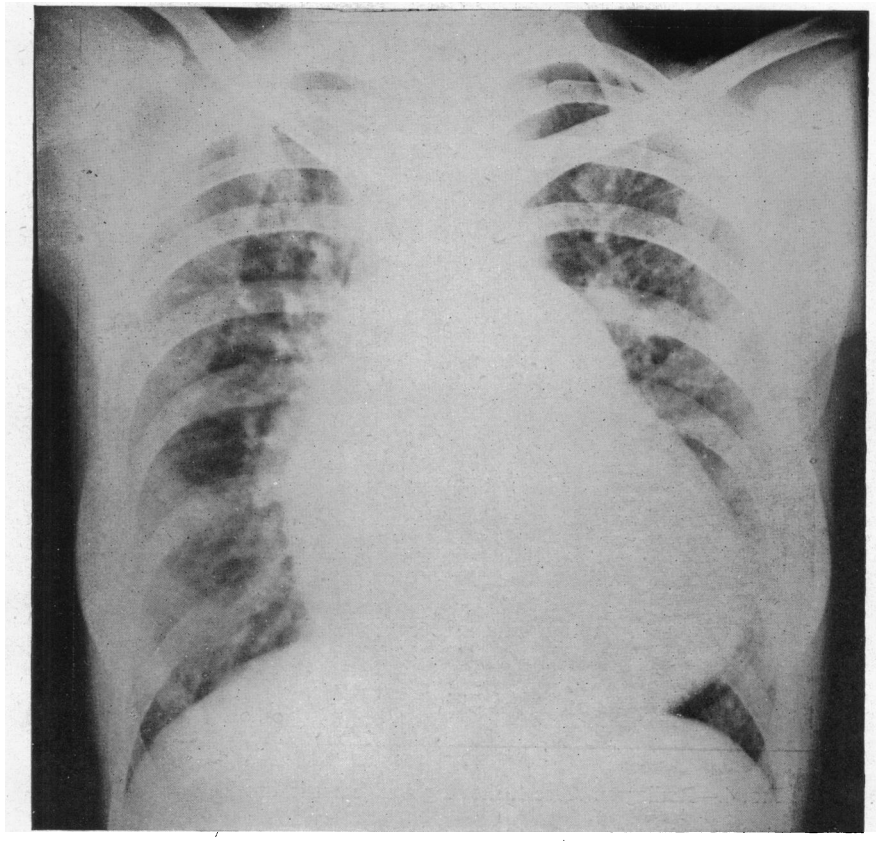

FIG. 5.—Radiogram Case 24, showing a globular heart with general enlargement, but no rib notching.

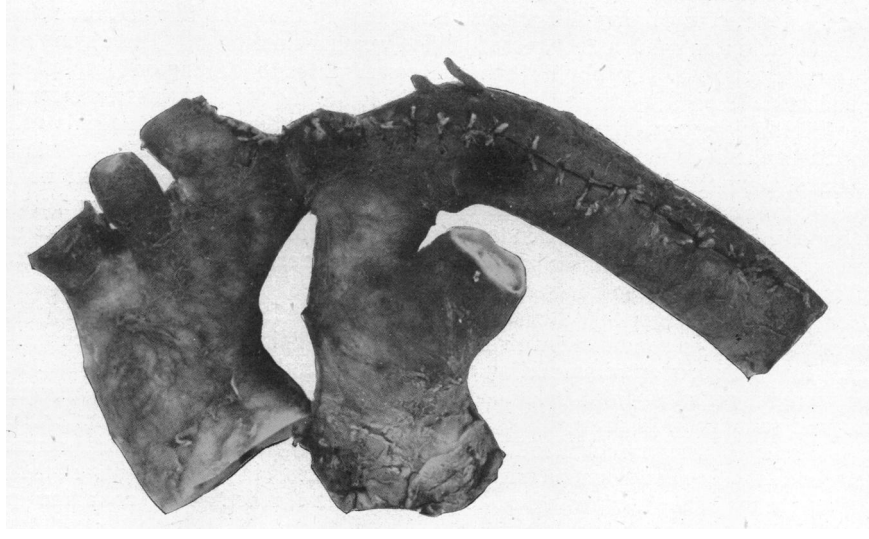

FIG. 6.—Autopsy specimen Case 24. Aorta, pulmonary artery, and patent ductus arteriosus.

hypertrophied but their cavities were not dilated. The right auricle was hypertrophied but the left auricle appeared normal. The ductus arteriosus was widely patent (Fig. 6). The diameter of the aorta at its origin was normal, but at the point of coarctation between the left subclavian and the ductus it would barely admit a pencil.

Coarctation had not been suspected in this case, though the peculiar paræsthesia in the legs should have aroused our suspicion. Since then in every case of patent ductus I have looked for evidence of coarctation, but hitherto I have found it only once.

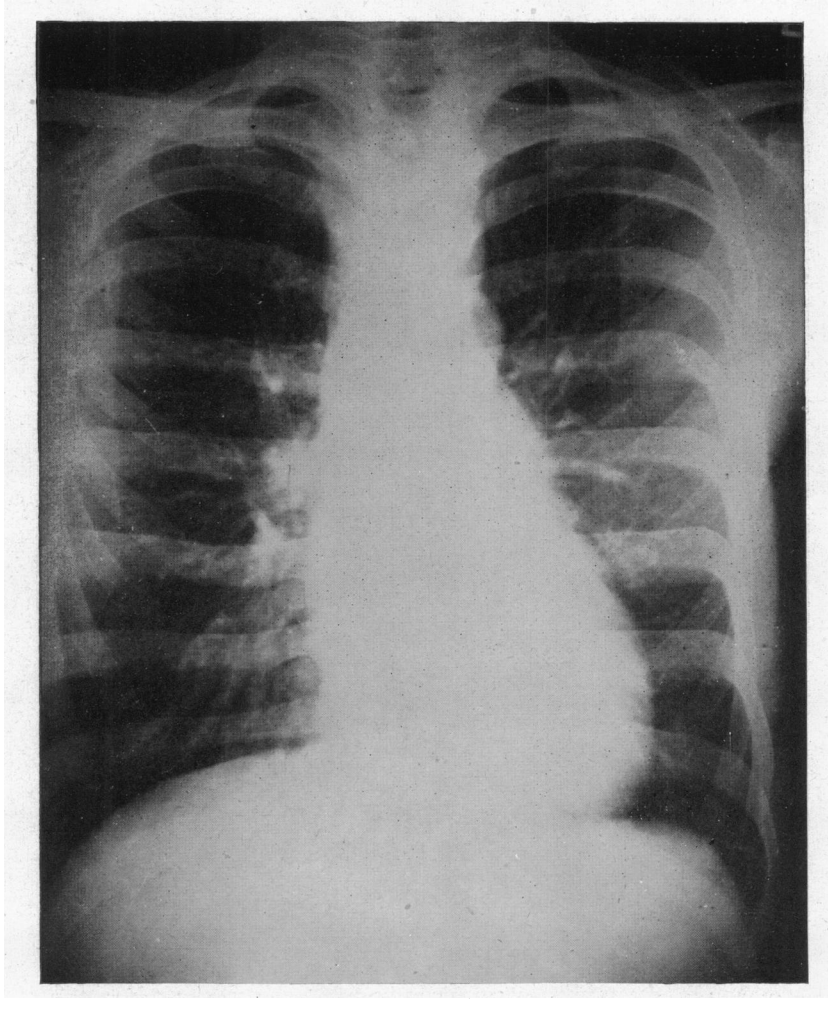

FIG. 7.—Radiogram Case 23 with coarctation and patent ductus arteriosus.

Case 23. A municipal civil servant, aged 27, consulted me because he had been rejected for another post on account of a cardiac murmur. It was the typical machinery murmur of patent ductus. The blood pressure in the arm was 165/80, but the systolic pressure in the dorsalis pedis was only 115. I, therefore, suspected the possibility of coarctation. There were no pulsating arteries on the back or front of the chest and radiography showed no rib notching, but it did show a double prominence in the region of the aortic knuckle (Fig. 7) and on screening it was obvious that the upper knob pulsated violently whereas the pulsation in the lower knob was feeble, the former being proximal and the latter distal to the coarctation.

Case 18. A boy, aged 11, was seen in consultation in September 1936. He had been a promising

athlete for his age and very fit until taken ill at school in July with a pain in the left thigh. Three weeks later he was admitted to a nursing home with a swinging temperature and an enlarged spleen. From August 10 until I saw him the temperature had been normal, but on September 8 he complained of a pain in the left chest and his pulse rate increased from 80 to 116 and subsequently remained rapid.

He had a systolic murmur both at the apex and at the base of the heart. The murmur was also clearly audible on the back of the chest. The blood pressure was 140/90. The spleen was easily palpable and he was considered to be suffering from subacute infective endocarditis. Progress was satisfactory till May 1937 when again, after a motor ride, his temperature rose to 99·4° F. and for a few days he had epistaxis. Subsequently, from time to time, he had pains in the chest, foot, shoulder, and other places, each associated with a rise of temperature.

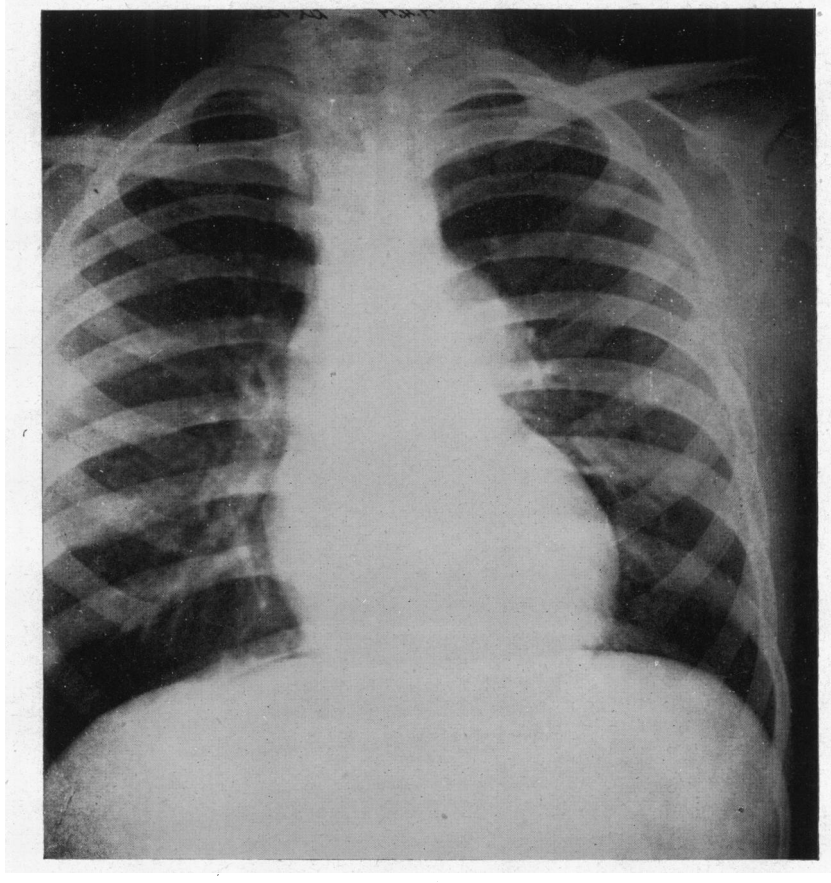

FIG. 8.—Radiogram Case 18, with coarctation and patent ductus arteriosus (August 20, 1937).

He was admitted to my ward on August 11, 1937, and it was then found that no pulsation could be felt in the femoral arteries. X-ray examination of the chest (Fig. 8) by Dr. Twining showed enlargement of the blind end of the descending aorta and enlargement of the pulmonary conus, but no rib notching. Dr. Twining suggested that the radiograms probably indicated coarctation of the aorta associated with patent ductus. Blood culture yielded a profuse growth of *Streptococcus viridans*. This was repeated and confirmed. He was treated with prontosil, but without benefit. On September 18 he developed a dry cough and the air entry in the left lung was found to be diminished. X-ray examination on September 25 showed partial collapse of the left lower lobe and on October 13 complete collapse of the left lung (Fig. 9). The pulmonary complications in this case suggested that the infection involved the ductus as well as the aortic valve.

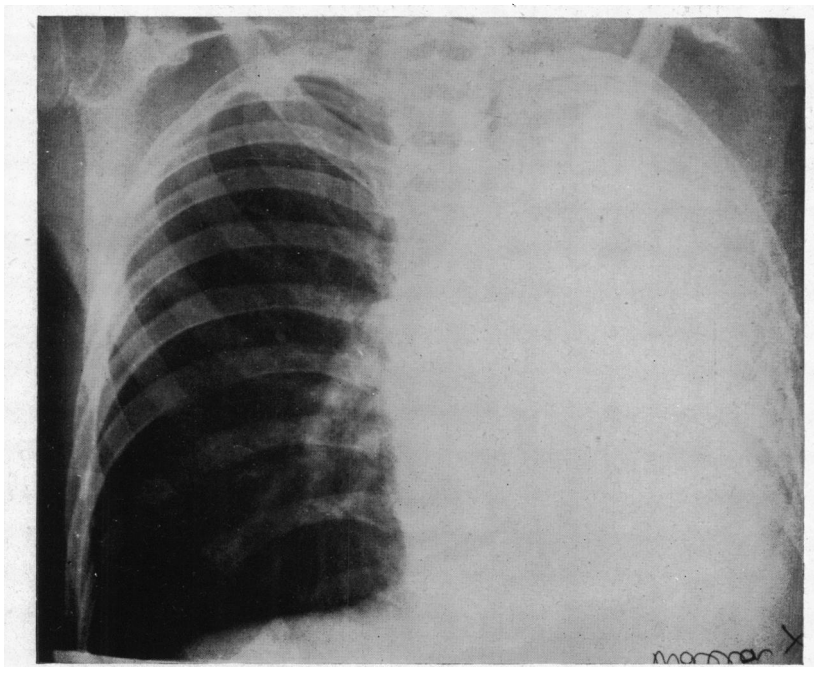

FIG. 9.—Radiogram Case 18, showing collapse of left lung (October 13, 1937).

PROGNOSIS

The number of cases in my series is too small to enable me to draw any firm conclusions regarding prognosis, but certain general deductions appear to be justified. In May, 1945, 10 of my patients were over 30, while 16 were under 30 years of age. Of the latter 3 have been lost sight of. In the older group 4 are dead, in the younger group 6. Of the 4 fatal cases in the older group 3 lived to over 50 years of age and in 2 of these death was not attributable to the coarctation, 1 died of carcinoma of the stomach and the other of lobar pneumonia. Of the remaining 6 patients in this group 5 are not merely alive but free from symptoms. One woman (Case 26) has passed safely through two pregnancies, one man (Case 4) is in a clerical post, and the other 4 all indulge in strenuous physical work. Symptoms dated from childhood in 4 of the 6 fatal cases in the younger group.

It would appear, therefore, that patients whose symptoms date from childhood are unlikely to reach the age of 30, whereas in those who are free from symptoms until the age of 30 the further expectation of life is much more favourable: they have passed safely through the hazards of the third decade.

To the two groups considered above there must be added a third with which I am not concerned in this paper, since young children do not figure in my series. Sixteen of the 28 patients in Evans's (1933) series died in infancy and in 15 of these the coarctation was associated with a patent ductus (see Table I).

Of 82 cases of coarctation collected by Abbott, in which death occurred within the first year, 50 exhibited some major congenital anomaly such as bilocular or trilocular heart, transposition of the arterial trunks, or pulmonary atresia. In the great majority of cases in which death occurs in infancy the coarctation is of the " infantile " type. These cases are of more interest to the embryologist than to the clinician.

It is interesting to compare the expectation of life in my series with that in the series of cases collected by Maude Abbott. Of her 200 cases, 103 (51 per cent) died before the age of 30 and a further 45, making a total of 148 (74 per cent) before the age of 40. In round figures, therefore, half her cases died before 30 and another quarter before 40. Thus between the

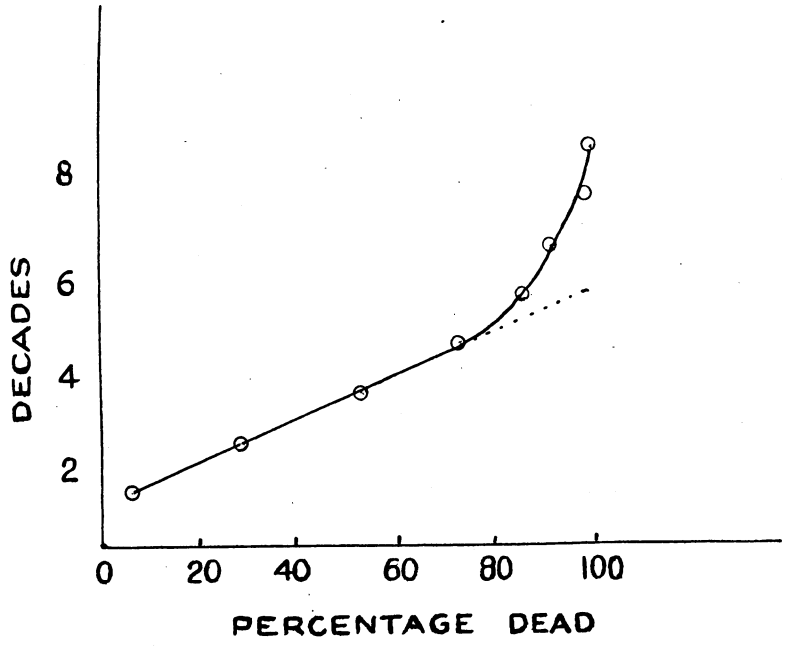

FIG. 10.—Graph showing age and mortality of coarctation of the aorta from Maude Abbott's series.

ages of 10 and 40 there is practically a linear relation between age and death-rate (Fig. 10). Had this been continued after 40, as shown by the interrupted line, all Abbott's cases would have been dead by the age of 50; but after 40, the death-rate tends to fall off, for in patients who reach 40 the coarctation often causes little serious disability, survival for a considerable period of years is not unlikely, and death not infrequently is due to some cause unconnected with the coarctation.

Blackford's figures are very similar to Abbott's. Of 180 cases over 5 years of age, 76 (42 per cent) died between the ages of 16 and 30 and 36 (20 per cent) between 30 and 40. Blackford states that many of these were young athletes who died during or shortly after exercise.

Cause of Death

The cause of death in the 10 fatal cases in my series is shown in Table VIII. Four died of congestive heart failure, 2 of subacute infective endocarditis, and 1 of each of the following conditions, septicæmia, subarachnoid hæmorrhage, carcinoma of the stomach, and lobar pneumonia.

Deaths from congestive heart failure occurred at the ages of 11, 23, 36, and 52. The other fatal cases fall in two well-defined age groups. The three patients who died of cardiovascular causes and the one who died of septicæmia were all under 30, whereas the two who died of

TABLE VIII

CAUSE OF DEATH (BRAMWELL'S SERIES)

Case No.	Age at death	Cause of death	Remarks
1	11	Congestive heart failure	Subaortic stenosis
18	12	Infective endocarditis	Patent ductus
2	23	Infective endocarditis	
15	23	Septicæmia	
24	23	Congestive heart failure	Patent ductus
8	27	Subarachnoid hæmorrhage	
9	36	Congestive heart failure	Two pregnancies Auricular fibrillation
3	52	Congestive heart failure	
5	52	Carcinoma stomach	
6	62	Lobar pneumonia	Auricular fibrillation

causes unconnected with their coarctation were both over 50. This suggests that in patients who survive the hazards of the first three decades the chief danger of coarctation is congestive heart failure.

Subacute infective endocarditis is a well-known complication of congenital heart disease, but the risk of subarachnoid hæmorrhage from rupture of a cerebral aneurysm has not received adequate recognition. Abbott believed that, in young subjects without history of trauma or infection, spontaneous subarachnoid cerebral hæmorrhage was usually due to a ruptured cerebral aneurysm of congenital origin. This was shown at autopsy to be the cause of death in 5 of her cases and she pointed out that the early age of the patients, the large collateral circulation, and the history of repeated small cerebral hæmorrhages suggested that the same was probably true of the other 13 cases of spontaneous cerebral hæmorrhage in which no aneurysm was actually demonstrated.

TABLE IX

CAUSE OF DEATH (ABBOTT AND BLACKFORD)

	Abbott	Blackford
Rupture of heart or aorta	44	38
Myocytic endarteritis of aorta	10	—
Cerebral lesion	24	25
Gradual heart failure	60	68
Sudden heart failure	17	16
Total deaths from cardiovascular causes	155	147
Total number of cases in series	200	196

Maude Abbott and Blackford both found that death was due to cardiovascular causes in over 70 per cent of the cases which they analysed (Table IX). My small series differs from those of Abbott and Blackford in that in no case was death due to rupture of the aorta, a complication that proved fatal in about 20 per cent of their cases. Three of Lewis's cases (Table X) died suddenly at the age of 31, 42, and 49, respectively, but in none of these was the cause of death determined, although in one a post-mortem examination was performed.

An associated congenital abnormality that appears important as a predisposing cause of death is a bicuspid aortic valve. This abnormality not only renders the patient more liable to subacute infective endocarditis but is frequently associated with a congenital weakness of

the aortic wall. A bicuspid aortic valve was present in 22 per cent of the cases in Abbott's series and in over 50 per cent of those in which death was due to spontaneous rupture of the aorta.

An aortic diastolic murmur was detected in only 4 of my patients (Cases 1, 3, 6, and 15). Perlman, on the other hand, reported a diastolic murmur in 10 of his 13 cases. This very high incidence is difficult to explain unless he applied the term " diastolic " to what other workers call a " late systolic " murmur.

Hazards of Coarctation

Bonnet (1903) maintained that fully compensated obstruction of the thoracic aorta made no extra demand on the cardiac reserve, and that hypertrophy of the heart was never present in uncomplicated cases. Lewis (1933) held the view that prolonged overwork in itself never led to heart failure. Abbott (1928) on the other hand believed that uncomplicated coarctation did entail cardiac strain which might by itself lead to death from failing compensation. She quotes a large number of observations in support of this hypothesis, and she found that cardiac hypertrophy was present in 75 per cent of the 200 cases she reviewed.

A detailed statement concerning cardiac enlargement in my own cases will be given in a later paper dealing with the radiology of coarctation. Here let it suffice to say that gross cardiac enlargement was present in only 4 of my 26 cases: in 3 of these (Cases 1, 3, and 6) it was associated with aortic incompetence and in the fourth (Case 24) with a freely patent ductus.

Case 2 is of particular interest in that we were able to compare the cardio-thoracic ratio at the age of 11 and 21. No alteration had occurred. Lewis (1933) reported little if any increase in heart size in those of his cases that were under observation for several years. These were older men: in my case there was no relative increase in heart size over the period of active growth.

My own observations lead me to believe that coarctation of the aorta does impose an additional load on the heart which entails a corresponding reduction in the cardiac reserve, but that, in the adult type of coarctation, this alone is not sufficient seriously to embarrass the heart during the first half of life, nor to render it incapable of meeting its everyday commitments. When coarctation is not complicated by the presence of some other heart lesion, symptoms of circulatory insufficiency are not likely to supervene unless the heart muscle be weakened by intercurrent infection, or an additional burden be imposed upon the heart by severe physical strain, in which case a progressive deterioration in health may ensue.

Not infrequently patients date their symptoms from some intercurrent infection which appears permanently to have upset the equilibrium. This is clearly illustrated by Lewis's series of cases of coarctation amongst pensioners from the First World War. These men had been invalided out of the army on account of cardiovascular symptoms and it is interesting to note the various conditions that were responsible for their breakdown. In three (Table X) symptoms of circulatory insufficiency developed following an attack of malaria. One (Case 3) developed symptoms at the age of 19 following an attack of P.U.O. which from Lewis's description sounds like trench fever. In a fifth (Case 6) a man of 27, who had served three years in the army, symptoms followed a foot infection. Thus in 5 of Lewis's 8 cases the onset of symptoms was attributable to intercurrent infection. There is, however, little evidence that a progressive cardiovascular deterioration occurred as the result of such infection since all five men survived for more than 10 and one for 24 years. This patient was 63 at the time Lewis's paper was published and subsequently attained the age of 73.

I am indebted to Dr. E. E. Pochin for the further history of this remarkable case, who was under treatment by Sir Thomas Lewis in October 1942 with increasing breathlessness over the

TABLE X

LEWIS'S CASES FROM THE 1914–18 WAR

Case	Predisposing cause	Age first symptom	Remarks	Duration (years)	Age at death	Cause of death
1	3 months' training ..	19	Previously active	12	31	" Sudden "
6	Foot infection ..	27	3 years' service	15	42	" Sudden "
7	Malaria	33		10	43	Infective endocarditis C.H.F. (P.M.)
8	After 4 months' France	41	Previously reserve	8	49	" Sudden " (P.M.)
4	Cycle accident ..	40	Detached retina	12	52	C.H.F. auricular fibrillation
5	Malaria	49	Rejoined 1914 aged 47	12	61	Acute pneumonia
2	Malaria	49	Enlisted 46 R.F.A. 3 yrs. foreign service	14+	—	Living 63*
3	P.U.O.	19	14 months front line, football	17+	—	Living 36

* Died 1944, aged 73.

previous twelve months and with several attacks of loss of consciousness during the preceding few weeks. He then could not walk more than 200 yards without having to stop owing to breathlessness and was made breathless by one flight of stairs. The attacks of unconsciousness occurred shortly after waking and were continued in hospital where he was found to have heart block. The degree of block was variable, some records showing a long P–R interval with a 1:1 ventricular response; most showing 2:1 block with a ventricular rate of about 44 with some failures of ventricular response and some extrasystoles; while some records showed complete heart block.

He had at this time 4 cm. of venous congestion, the cardiac apex beat was 15 cm. to the left in the fifth space, the right border of dullness being 4 cm. to the right of the mid-line. The blood pressure was 200/120 in the arms and 80/60 in the legs. On treatment with ephedrine his Stokes-Adams attacks ceased and his heart rhythm became regular at a rate of about 80, but he still showed alternation of the pulse and gallop rhythm. He was seen at intervals until October 1944 when he died, out of hospital. No details of his form of death are available, but a post-mortem was obtained and the specimen is now in the Pathological Department of University College Hospital.

Severe physical strain is another hazard to which these patients are liable to be exposed, for as Abbott points out the majority are males and many are of an athletic, muscular build. Blackford (1928) states that in many cases sudden death from rupture of the aorta has occurred during or immediately after athletic exertion.

Pregnancy and Coarctation

A correct appreciation of the dangers of childbirth in women with coarctation is important, since grave complications are liable to develop unexpectedly during the second stage of labour. Two* of the patients in my series have been under observation during pregnancy.

Case 26. A woman aged 37 was referred to my clinic in the fourth month of her second pregnancy on account of a history of mitral stenosis. She was delivered by Cæsarean section in August 1945 and when last seen in August 1946 was well except for slight breathlessness on exertion. Up to the age of 20, when she first consulted a doctor on account of breathlessness when cycling and œdema of

* Two other cases seen since May 1945 have been successfully delivered by Cæsarean section.

the ankles, she had been perfectly fit. A heart murmur was discovered at that time. This was wrongly attributed to mitral stenosis and she was accordingly kept in bed for six weeks. Subsequently, until her marriage at the age of 24, she worked as a machinist and lost little time off work through ill health, but strenuous exertion made her breathless. Convalescence following her first confinement—a forceps delivery at the age of 29—was prolonged and she was in bed for six weeks, but from then onwards she enjoyed good health until she again became pregnant. Apart from her illness at the age of 20, which appeared to be attributable to over-exertion, this woman had little in the way of symptoms except during her two pregnancies.

Case 9.—A primigravida, aged 31, was admitted to St. Mary's Hospital, Manchester, with a view of Cæsarean section for contracted pelvis, and was referred to my clinic in March 1933 on account of high blood pressure. She had been unaware of any heart trouble up to the fourth month of pregnancy when she had noticed some shortness of breath and palpitation. For sixteen years before her marriage at the age of 29, she had worked hard on a farm where her daily routine included heavy lifting and scrubbing.

She was successfully delivered by Cæsarean section in April 1933. Six weeks later she developed a severe hypochromic anæmia which responded well to treatment and she then remained free from symptoms until November 1935 when she began to complain of lassitude, palpitation, and præcordial pain on exertion. On examination it was found that she was again pregnant and had developed auricular fibrillation. In May 1936 she had a complete abortion from which she made a good recovery, but subsequently she developed congestive heart failure from which she died in May 1938. This case has already been fully reported (Bramwell and Longson, 1938).

A case reported by Walker (1943) presented some similarity to the one described above. She also was a farmer's wife, 31 years of age, but before marriage her activities had been considerably restricted. She remained fit throughout pregnancy and tolerated the first stage of labour well, but became acutely distressed and breathless following rupture of the membranes. She was delivered by forceps and convalescence was uneventful.

The alarming complications that may develop unexpectedly during labour are illustrated by a case reported by Billinghurst (1943). His patient, a healthy and athletic primipara of 23, who had no symptoms during pregnancy, fainted at the onset of the second stage of labour and died suddenly the day after delivery from rupture of the aorta.

TABLE XI

JENSEN'S CASES OF COARCTATION ASSOCIATED WITH PREGNANCY

Author	Date	Age	No. of children	Remarks
Leudet	1858	37	4	Died from rupture of aneurysm just beyond coarctation
Kreigh	1878	28	2	Died following stroke in 9th month of her 3rd pregnancy
Fawcett ..	1905	45	9	Died of congestive heart failure 14 days after induction of premature labour
Abbott	1915	38	1	Died of congestive heart failure some years after confinement
Katz	1921	25	1	Died on rupture of the aorta near the end of 2nd pregnancy
Strassmann ..	1922	56	7	Found dead in bed
Strayhorn ..	1937	26	0	Successfully delivered by Cæsarean section

In 4 of the 7 cases collected by Jensen (1938) (Table XI) death occurred either during pregnancy or shortly after delivery from causes directly related to the coarctation : but one of these women had survived eight previous pregnancies and another lived to the age of 56 after having had seven children.

Summary and Conclusions

Twenty-six cases of coarctation of the aorta are reported. In 20 the diagnosis was based on clinical evidence, in 5 it was made by the radiologist, and in 1 the lesion was discovered post-mortem.

The classification, clinical manifestations, and prognosis of the condition are discussed and the following conclusions are reached.

Coarctation of the aorta is apt to escape recognition, unless the possibility of the condition be continually kept in mind. Many of Lewis's cases were under observation for years before they were correctly diagnosed. When associated with a patent ductus or with aortic incompetence, coarctation is especially liable to be overlooked.

Clinically the diagnosis is based on the finding of high systolic blood pressure in the upper limbs associated with low pressure in the lower limbs, pulsating arteries on the back or front of the chest, excessive arterial pulsation at the root of the neck, and a late systolic murmur of abnormal distribution. The most important radiological manifestations are rib notching and occasionally a double aortic knuckle. Direct demonstration of the coarctation is rarely possible. Diagnosis is most difficult when the ductus is patent, for then there may be no collateral circulation and no notching of the ribs.

In many cases, symptoms first appear following some intercurrent infection or physical strain. Dyspnœa, palpitation, and fatigue are the most common complaints. Pain in the trunk and arms may be due to pressure on nerves by the dilated arteries participating in the collateral circulation. The spinal anastomosis is of importance in this connection.

Despite a deceptively good exercise tolerance, a sudden strain may prove fatal even in the prime of life: these patients are living dangerously. It is accordingly wise to limit their activity. Women with coarctation should be delivered by Cæsarean section.

In coarctation of the aorta fatal complications are so liable to develop unexpectedly that prognosis is extremely uncertain: a patient who appears to be in robust health one day may be dead the next.

Patients in whom symptoms date from childhood rarely live to 30, whereas those who surmount the hazards of the first three decades may suffer little disability from their coarctation. The efficiency with which the circulation may be maintained year after year, in spite of the increased load due to high blood pressure, is remarkable.

I am indebted to Dr. A. Morgan Jones for his help in the investigation of these cases, to Dr. E. Duff Gray and the late Dr. E. W. Twining for the radiograms, to Dr. W. Susman for the pathological observations in Case 24, to Dr. E. E. Pochin for particulars of Lewis's case, to Dr. William Evans and the Oxford University Press for permission to reproduce Fig. 1-3, and to other colleagues for referring cases to me.

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APPENDIX

Two patients in this series who were alive in May 1945 have since died. Their subsequent history and the post-mortem findings were as follows.

Case 13. This man was admitted to the Bolton Royal Infirmary on 15/10/45 suffering from subarachnoid hæmorrhage from which he made an uneventful recovery. He was readmitted on 9/12/46 following a severe hæmoptysis. His blood pressure then was 130/70. During the ensuing week three further hæmoptyses occurred, and he died on 16/12/46.

I am indebted to Dr. Horace Jackson for the further clinical history in this case and for the following report on the post-mortem examination. There was a cavity filled with fresh blood clot in the lower lobe of the right lung and the surrounding tissue was saturated with blood. The hæmorrhage appeared to have resulted from rupture of a fairly large vessel, but there was no clear evidence of an aneurysm. There was no tuberculosis. Just distal to the origin of the left subclavian artery, at the point of insertion of the ductus, which was closed, the aorta was narrowed, but not completely occluded (Fig. 11 on p. 126). It admitted a probe 0·2 cm. in diameter. The internal mammary vessels were very large. Examination of the brain revealed an aneurysm about 1 cm. in diameter on the circle of Willis. The surrounding brain showed staining from an old hæmorrhage—probably that from which the patient suffered in 1945.

Case 20. This young man kept well until April 1946 when, after rowing, he had an attack of sweating, and for three days was febrile, complaining of pain in the right thigh and right foot. Four weeks later he had another febrile attack, again after rowing. He was then seen by Dr. D. Rhodes Allison who diagnosed subacute bacterial endocarditis and admitted him to a private ward at the Manchester Royal Infirmary on 7/6/46. I am indebted to Dr. Allison for the opportunity of seeing this patient at that time and for the further particulars of his illness. He was treated with penicillin and progress was satisfactory until 27/6/46 when he complained of pain in the chest. Two days later he vomited copiously and from that time his condition steadily deteriorated. On 2/7/46 he collapsed suddenly and died.

Post-mortem examination by Dr. F. A. Langley showed a narrowing of the aorta just proximal to the insertion of the ductus arteriosus which was closed, the lumen of the aorta at this point being less than 0·5 cm. in diameter, whereas that of the descending aorta was 1·0 cm. in diameter (Fig. 12 on p. 127.) Beyond the coarctation, for a distance of 3 cm. the wall of the aorta was much thickened and roughened and small vegetations were present. The internal mammary artery and its branches were very large. They did not directly anastomose with the deep epigastric artery but broke up into small muscular vessels in the rectus abdominis just above the umbilicus. The heart weighed 700 g., the left ventricle being greatly dilated. The interventricular septum was displaced to the right reducing thereby the capacity of the body of the right ventricle (Bernheim's syndrome). The conus of the right ventricle was dilated. The right auricle was dilated and its wall somewhat thickened. The left auricle appeared normal. The aortic valve was bicuspid, both coronary arteries arising from the anterior sinus of Valsalva; the anterior cusp was thickened and on it there were a few small vegetations; the posterior cusp had a large vegetation covering its ventricular surface. The coronary arteries appeared normal.

I am indebted to the Department of Pathology of the University of Manchester for the photographs of the post-mortem specimens.

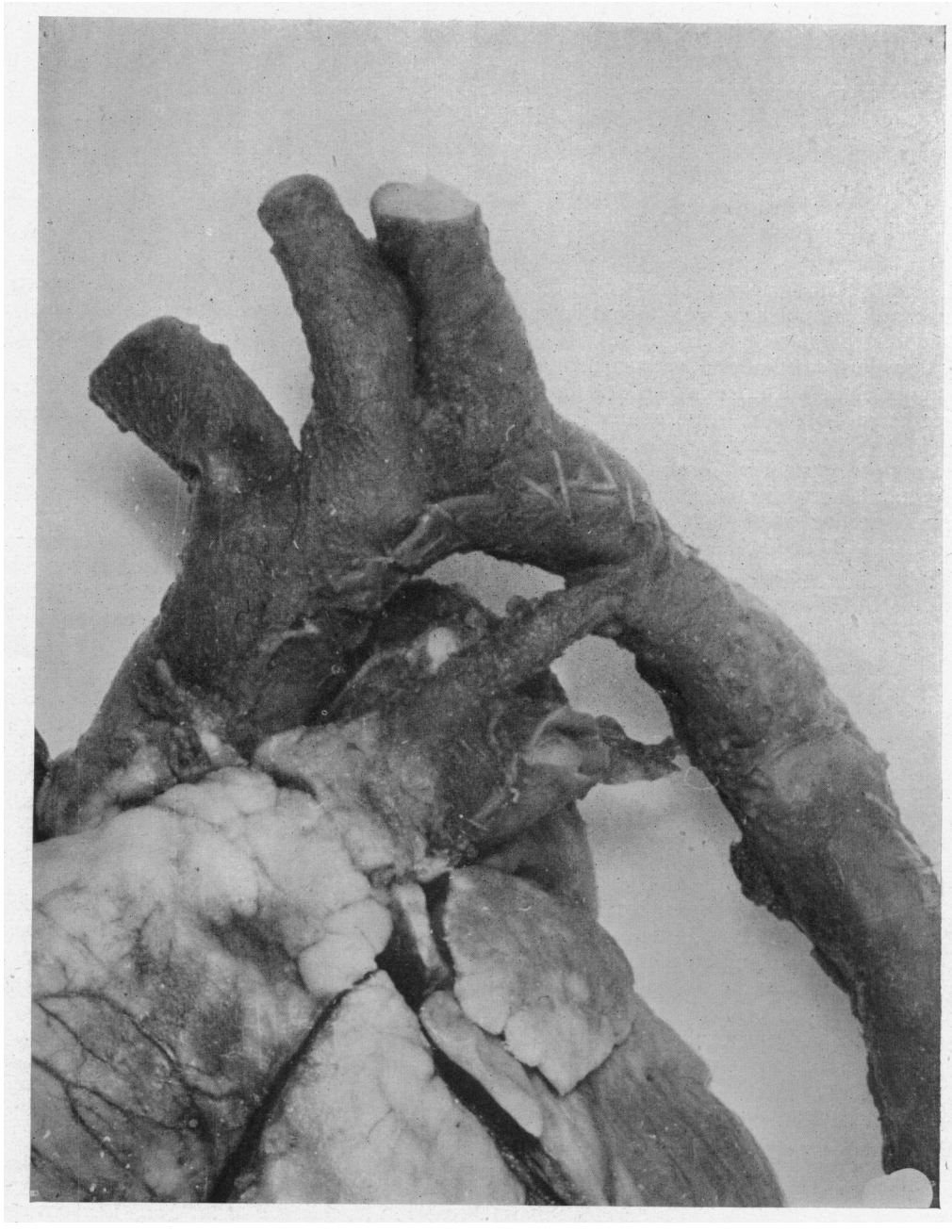

FIG. 11.—Specimen of coarctation from Case 13.

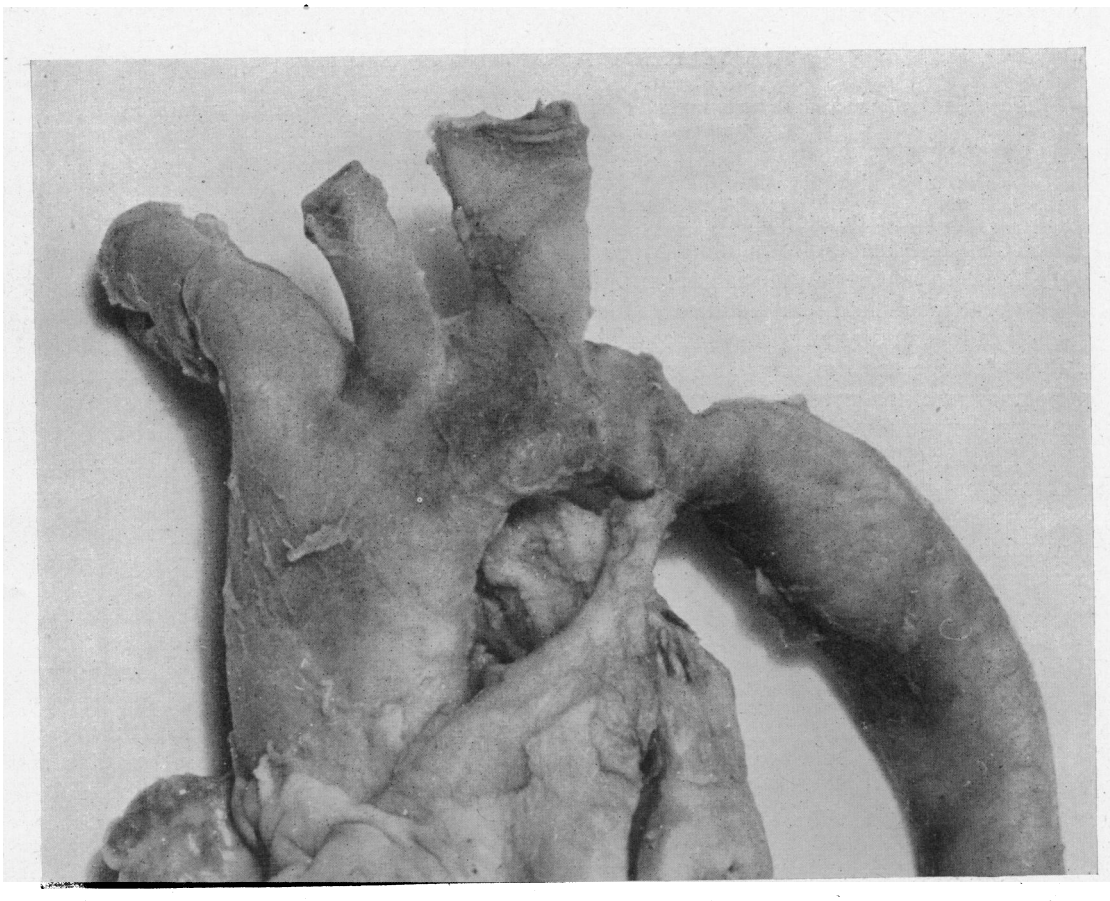

FIG. 12.—Specimen of coarctation from Case 20.