

RETINAL CHANGES IN COARCTATION OF THE AORTA*

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COARCTATION of the aorta (stenosis of the isthmus) is a congenital narrowing or occlusion of the aorta immediately below its junction with the subclavian artery at the orifice of the ductus arteriosus. The most characteristic symptom of this unusual disease is hypertension in the upper half of the body and relatively low blood pressure in the lower half (in normal persons the blood pressure is somewhat higher in the legs than in the arms). The blood supply to the lower part of the body takes place through numerous collaterals from the carotids and subclavians. Notching of the ribs is frequently visible radiologically. There is usually relative freedom from symptoms up to the age of about 15, when a number of hypertensive and cardiac symptoms appear. Approximately 50 per cent. of persons with this disease are stated to survive until the age of 40, and only 20 per cent. to the age of fifty.

Crafoord's operations have caused an unusually large number of cases of this uncommon disorder to be assembled in Stockholm. The patients come not only from various parts of Sweden but also from other countries in Europe.

Before an operation is considered, the patients undergo a thorough examination in Professor Nylin's cardiological department at Södersjukhuset. As in other cases of hypertension, these patients are sent as a matter of routine for examination to the eye department. As soon as I had seen a few such cases, it became evident that the retinal picture in coarctation of the aorta is often somewhat characteristic, the principal feature being pronounced tortuosity of the arteries. In a preliminary report at the Annual Meeting of the Swedish Ophthalmological Society (1948), I gave an account of my findings in seventeen cases. The aforementioned characteristic picture was present in ten of these patients; I therefore expressed the opinion that it should be possible—at least in more pronounced cases—to distinguish between coarctation of the aorta and other cases of juvenile hypertension by means of ophthalmoscopy.

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I have been able to find relatively few reports in the literature of retinal changes in coarctation of the aorta. For example, no mention is made of the fundi in the survey of thirteen cases given by Perlman (1944), in the report of 26 cases by Bramwell (1947), the report of fifteen cases by Campbell and Suzman (1947), or in the report of 26 cases by Newman (1948). Kartagener and Zimmer (1940) stated that the retinal vessels were normal in their cases, and Ask-Upmark (1942) reported a normal retinal picture in one of his three cases.

In a case reported by Lichtenberg and Gallagher (1933), mention is made of marked pulsations in both the retinal arteries and veins, and Nicolson (1940) stated that, in his cases, the retinal vessels were tortuous and pulsating. Rhodes and Durbin (1942) described three cases of coarctation of the aorta in children, in one of whom the retinal arteries appeared narrow and more tortuous than normal, while the veins were not unusual. They added:

in several cases in children the retinal arteries have been described as constricted and tortuous.

In four of the five cases described by Schwartz (1946), it was mentioned that the retinal arteries were tortuous or possibly narrowed as well, whereas in one case these vessels were normal. All four abnormal cases were diagnosed as "retinal arteriosclerosis". In a symposium on coarctation of the aorta from the Mayo Clinic, Christensen and Hines (1948) discussed the clinical picture on the basis of 96 cases, on eighty of which ophthalmoscopy had been performed. A normal picture was found in only 22 per cent.; generalized narrowing of the retinal arterioles was frequent; higher grades of localized narrowing occurred in some cases. Of interest is the fact that none of the eighty patients had severe hypertensive retinopathy. No mention was made of tortuosity of the arteries.

Lundar (1948) described a case in which the retinal arteries were extremely narrow and tortuous with no variations in calibre. The veins were normal and there were no haemorrhages or retinal patches. Connelly and Gibson (1949) reported on a patient who had been under observation for 14 years, and showed pronounced tortuosity of the retinal arteries. They considered this to be a vascular anomaly and not an indication of sclerosis. They stressed that hypertension in coarctation of the aorta and essential hypertension are not caused by the same mechanisms.

In a survey of the diagnostic methods in coarctation of the aorta, Proudfit and Ernstene (1949) made no mention of examination of the retina.

Tortuosity of the retinal arteries is thus mentioned in many of the cases reported in the literature. In no instance, however, has it been stressed that the retinal changes in this disease are in any way characteristic.

PRESENT INVESTIGATION

Between 1947 and 1950, forty undoubted cases of coarctation of the aorta were sent from the cardiological department for ophthalmological examination, and I soon noticed that tortuosity of the arteries was characteristic of many such cases. It was often strikingly regular, and, in some less marked cases, it was confined to certain arterial branches, whereas in others it could be seen in the majority of the branches. Initially, the tortuosity appeared to me to be associated with regular, undulating changes in the width of the artery, and with regular changes in the light reflexes. The examination was always made with Gullstrand's binocular ophthalmoscope, in which a stereoscopic picture of the fundus is consistently obtained. This soon revealed that these changes in the arterial width and in the light reflexes were, in all probability, only apparent. It was possible to see clearly how the arteries were often twisted in the shape of a corkscrew, and were, in some places, somewhat embedded in the retinal tissue, whereas other sections of them lay more superficially. This explains the regular changes in the light reflexes and the apparent variations in calibre. (In looking at a spiral metal thread, the changes in the light reflexes are noticed immediately and also—particularly under certain conditions of illumination—apparent variations in width.)

More or less pronounced tortuosity of the retinal vessels may occur in normal cases, although it is then, as a rule, mainly localized to one plane and is infrequently found to be in the shape of a corkscrew. Moreover, in normal cases, it is often present in both the arteries and the veins. In coarctation of the aorta, on the other hand, the veins are seldom tortuous.

In the Table overleaf the patients are listed according to age and numbered in the order in which they were sent for examination.

The Table shows that more or less pronounced tortuosity was present in 24 of the forty cases; furthermore, it is evident that the corkscrew-like formation increases in frequency with advancing age. It was present in eleven out of the 24 patients below the age of 25, as against thirteen out of the sixteen aged 25 years or more. This change in the retinal arteries may therefore be considered as fairly characteristic of coarctation of the aorta, particularly in older patients. The changes are easily seen in marked cases, whereas they may readily be overlooked when they are slight, unless attention is particularly focused on their possible occurrence. Examination with a binocular ophthalmoscope is of especial value for the detection of the corkscrew formation. It may be added that it is often possible to observe very clearly swinging pulsations of the entire arterial branches.

I re-examined a few patients after they had been operated on. The retinal picture was then exactly the same as at the first examination one or two years earlier.

TABLE
 PATIENTS IN ORDER OF AGE SHOWING GRADES
 OF RETINAL CHANGES

Patient No.	Age (years)	Grade	Patient No.	Age (years)	Grade	Patient No.	Age (years)	Grade
35	8	.	16	20	+-	18	25	-
19	9	.	33	20	—	26	25	—
12	11	---	36	20	+	15	26	++
22	11	---	31	21	—	30	26	++
20	12	---	23	22	—	40	26	-
21	12	---	29	22	—	7	28	.
38	14	.	39	22	+	34	28	-
3	15	.	9	23	++	28	31	—
6	16	---	32	23	+	27	32	.
24	16	.	2	24	+	14	34	—
25	18	.	5	25	++	37	36	++
8	19	---	11	25	+	10	39	+
4	20	---	17	25	.	1	42	++
13	20	.						

· + marked corkscrew tortuosity. · slight but unmistakable changes. — no arterial change.

In a few cases there also appeared to be a generalized narrowing of the arteries. Local arterial constrictions—which are often found in essential hypertension—were, on the other hand, usually lacking. Slight changes of this kind or slight arterio-venous crossing signs were present only in a few instances. In no case did I find haemorrhages, white patches, or other retinal changes occurring in severe hypertensive retinopathy. In this disorder, tortuosity of the arteries, even in the form of a corkscrew, is sometimes found, particularly in the macular region. In such cases, however, other typical arterial changes are always present concurrently. In coarctation of the aorta, on the other hand, the arteries have an essentially normal appearance, with the exception of the tortuosity. Moreover, the tortuosity is not especially confined to the macular region.

It therefore appears evident that the retinal picture in coarctation of the aorta differs distinctly from that usually found in other forms of hypertension.

- (1) Corkscrew-shaped tortuosity of the arteries is common in the former disease, particularly in patients over the age of 20 or 25.
- (2) The retinal changes characteristic of hypertension of other origin are wholly or partly lacking in coarctation of the aorta.

It should therefore be possible—at any rate in certain cases—to differentiate between coarctation of the aorta and other forms of juvenile hypertension by means of a thorough examination of the fundi. Case 37 is a particularly good illustration of the fact that ophthalmological examination can contribute to a diagnosis of coarctation of the aorta.

Case 37. A 32-year-old woman, who was to be operated on for an ovarian cyst, was found to suffer from hypertension. She was therefore transferred from the gynaecological to the cardiological department for further examination. After the diagnosis of hypertension, she was sent for ophthalmological examination, which revealed not only small variations in the calibre of the arteries and slight arterio-venous crossing signs, but fairly pronounced corkscrew-like tortuosity of the arteries. For this reason, I suggested that it might be a case of coarctation of the aorta and not of essential hypertension. This tentative diagnosis was confirmed by measurements of the blood pressure in the legs and a further study of the radiograms.

Tortuosity of the arteries, often in corkscrew form, is common in the entire upper half of the body in coarctation of the aorta, becoming more pronounced with the years. Tortuosity of the retinal arteries is also more frequent with advancing age. It therefore appears probable that the tortuosity of the retinal arteries is only a partial symptom of generalized arterial tortuosity, which is a result of the abnormal mechanical conditions.

Hypertension in the upper half of the body is also, presumably, mainly due to mechanical factors. Some authors have assumed that it is caused by a decrease in the blood supply to the kidneys, but other writers have put forward strong arguments against this hypothesis. In coarctation of the aorta, the absence of marked retinal changes, other than tortuosity of the arteries, is, in my opinion, a strong indication that hypertension in this disease is of an entirely different nature from that found in other hypertensive disorders. In the latter, the condition of the finer arterial branches is probably of prime importance, both for the occurrence of hypertension and for the possible retinal changes. Conversely, in coarctation of the aorta, the arterial changes (tortuosity) are of a more secondary nature. It is probable that both the tortuosity of the arteries and the hypertension are mechanically conditioned by the increased resistance. The pathogenesis of other hypertensive disorders is certainly considerably more complicated.

SUMMARY

(1) An account is given of characteristic retinal changes found in 24 out of forty cases of coarctation of the aorta. The changes consist of more or less pronounced corkscrew-shaped tortuosity of the retinal arteries, most distinctly visible with Gullstrand's binocular ophthalmoscope. Other retinal changes usually present in hypertensive disorders of a different nature are almost entirely lacking.

(2) It is therefore suggested that, in juvenile hypertension, careful examination of the fundi can contribute to a diagnosis of coarctation of the aorta.

(3) The difference between the causative mechanism in hypertension in coarctation of the aorta and in other hypertensive disorders is discussed.

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