

RETINAL CHANGES ASSOCIATED WITH COARCTATION OF THE AORTA

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REVIEW

COARCTATION OF THE AORTA is a congenital malformation producing significant narrowing of the lumen in a short thoracic segment of the vessel. In most instances the narrowed segment is located between the origin of the left subclavian artery and a zone just distal to the aortic insertion of the ductus arteriosus. A generally accepted classification introduced by Bonnet (1) separates the anomaly into infantile and adult types. In the former, the zone of narrowing lies between the left subclavian and the ductus arteriosus; whereas in the adult type it is located at or immediately distal to the ductus arteriosus.

The incidence of coarctation of the aorta is estimated by Blackford (2) to be 1:1500 of the general population. He arrived at this figure by analysis of a series of 68,300 routine autopsy reports.

Although minor degrees of coarctation are not incompatible with a long and useful life, according to Gross (3) the average age of survival as a group is 35 years, including those individuals who died from incidental causes. The common causes of death from the disorder include rupture of the aorta, cardiac failure, intracranial hemorrhage, and bacterial endocarditis and aortitis.

For many years the condition received only cursory interest from the medical profession, but since the development of adequate surgical treatment during the past few years, there has been a revival of interest in the vascular anomaly. The importance of early diagnosis and treatment before serious cardiovascular damage has occurred is now recognized by all.

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Diagnostic features of the disease include a high arterial blood pressure in all branches of the aorta proximal to the narrowed segment, and a lower blood pressure in all branches peripheral to the stenosis. The pulse pressure tends to be wide in the upper compartment and narrow in the lower compartment. Characteristically, feeble or absent pulsations along with retardation and prolongation of the pulse wave are found in the lower extremities. Left ventricular hypertrophy and a well-developed collateral circulation can usually be demonstrated. Roentgenographic examination often discloses local erosion of the inferior aspect of the posterior portion of the ribs from enlarged and tortuous intercostal arteries, often giving a scalloped appearance to the rib margin. Retrograde aortograms allow visualization of the constricted segment and provide valuable information concerning the site and character of the coarctation. It is our belief that coarctation of the aorta also produces alterations in the retinal arterioles, and that recognition of these changes may aid in the diagnosis of the disease.

REVIEW OF LITERATURE

Granström (4), in a report published in 1951 on the retinal changes in 40 patients with coarctation of the aorta, first stressed the peculiar spiral tortuosity produced in the arterioles. He introduced the term "corkscrew tortuosity" and observed the change in 60 percent of 40 cases. In addition, he frequently observed ". . . very clearly swinging pulsations of the entire arterial branches." Generalized narrowing, localized constrictions and sclerosis were noted in a few cases. He examined a few patients one or two years after surgical treatment and found the retinal picture exactly the same as at the time of the initial examination.

In a symposium on coarctation of the aorta, Christensen and Hines (5) reviewed the clinical features of the disease in 96 cases. The fundi were examined in 80 patients and only 22 percent were reported as normal. The remainder presented varying degrees of generalized narrowing, localized narrowing, and sclerosis. None of the patients presented severe hypertensive retinopathy. Tortuosity or pulsation of the retinal arterioles was not mentioned.

Schwartz (6), reporting on five cases in 1946, described tortuosity and sclerosis in four patients and found normal retinal arterioles

in the other. Rhodes and Durbin (7) reported tortuosity and narrowing of the retinal arterioles in one of three children with coarctation of the aorta. In 1940 Nicolson (8) described tortuosity and pulsation of the retinal vessels in a case of coarctation. Connelly and Gibson (9) noted increased tortuosity of the retinal arterioles in their patient, but considered this to be a vascular anomaly since the veins were also tortuous. Marked pulsation of the retinal arterioles and veins was described in a single case by Lichtenberg and Gallagher (10) in 1933.

With the exception of Granström (4), none of the reports indicate that the retinal changes were considered in any way characteristic of coarctation of the aorta. In several surveys of large numbers of cases no reference is made regarding ophthalmoscopic examinations. These include Campbell and Suzman (11), 15 cases, 1947; Bramwell (12), 26 cases, 1947; Perlman (13), 13 cases, 1944; Newman (14), 23 cases, 1948. In 1949 Proudfit and Ernstine (15) reviewed the diagnosis of coarctation of the aorta, but made no reference to fundus examinations.

OBSERVATIONS

This report is based on eleven cases of coarctation of the aorta which have been examined during the past year. Eight of the cases had undergone successful surgical treatment before the initial examination. Two cases were examined before and after surgical treatment. One patient, the oldest in the group, was not subjected to surgery because of his age. Even though we were unable to examine the fundi of a majority of the patients prior to surgery, it is considered unlikely that any change occurred during the interim which would in any way alter the observations made at a later date. The patients who were examined prior to surgery presented exactly the same picture at later examinations. Granström (4) examined patients at intervals of one to two years following surgery and found no change in the fundus details.

The ages of the patients ranged from five years to 44 years and included six males and five females. All members of the group prior to surgery had hypertension in the upper compartment and normal to slightly low blood pressure in the lower compartment. Pulsations in the femoral arteries were feeble or absent and the

pulse wave was retarded and prolonged in all cases. All but one patient, Case 1, presented evidence of notching of the ribs and in all cases there was hypertrophy of the left ventricle. The coarcted segment and a well-developed collateral circulation were demonstrated by retrograde aortograms in each of the patients.

Seven of the eleven patients presented visible changes in the retinal arterioles. The changes consisted of:

- 1) a peculiar spiral or "corkscrew" tortuosity of the arterioles;
- 2) serpentine pulsation of the arterioles.

The tortuosity was marked and involved most of the arteriolar branches in four of the patients and was considered rather characteristic of coarctation of the aorta. Only one of these presented comparable tortuosity of the veins. In three other cases the tortuosity was less marked and confined largely to a few branch arterioles. These findings were considered suggestive but not characteristic of coarctation of the aorta.

Ten of the patients had slight to marked, ophthalmoscopically visible, serpentine pulsations in the arterioles. A notation regarding pulsation was not recorded in the remaining case.

Varying degrees of arteriolar sclerosis, generalized narrowing and localized constrictions were noted in six patients. These changes were closely related to the duration of the disease, the

RETINAL ARTERIOLAR CHANGES IN COARCTATION OF AORTA AND RELATION
TO HYPERTENSION AND AGE OF PATIENT

Case	Age	B/P Rt. arm	Spiral ^a Tortu- osity	Serpentine Pulsa- tions	General- ized Sclero- sis	Gener- alized Narrow- ing	Local- ized Constric- tions	Venous ^a Tortu- osity
1	5	160/90	0	Present	0	0	0	0
2	7	140/70	++	Present	0	0	0	0
3	8	144/92	0	Present	0	0	0	0
4	11	180/100	++	Present	0	0	0	0
5	18	140/90	++	Present	0	0	0	++
6	18	170/105	0	Present	Grade 1	Grade 1	0	0
7	19	190/100	+	Present	0	Grade 1	0	0
8	20	170/120	++	Present	Grade 1	Grade 2	Grade 1	0
9	31	190/100	+	Not Recorded	Grade 2	0	0	0
10	37	190/100	+	Present	Grade 1	0	0	+
11	44	210/116	0	Present	Grade 3	Grade 1	0	Occlusion of Central vein

^a ++ = Marked. + = Slight.

severity of the hypertension, and especially to the level of the diastolic pressure. No evidence of hypertensive retinopathy was discovered in any case, but the oldest patient presented a partial closure of the central vein in one eye.

The results of this study are summarized in the table. The data is arranged to show the frequency and type of vascular change and the relation between these changes, the severity of the hypertension and the age of the patient.

DISCUSSION

Tortuosity of the retinal vessels occurs rather frequently in healthy persons and is thought to be congenital in nature. Also, certain diseases, such as congenital heart disease, leukemia, and polycythemia, are accompanied by tortuosity of the retinal vessels. Usually, however, in such cases both sets of vessels are affected to a comparable degree and not infrequently the veins are more tortuous than the arterioles. In our cases of coarctation of the aorta, the tortuosity was confined largely to the arterioles. In only one patient with well-developed changes was the tortuosity of the veins comparable to that of the arterioles. Further, the tortuosity which we have observed in coarctation has a peculiar spiral or "corkscrew" appearance which is quite striking in the well-developed case. The bends in the vessels in the usual types of tortuosity tend to be from side to side and fairly well localized to one plane.

The factors which operate to produce tortuosity and pulsation in the retinal arterioles have been thoroughly reviewed by Friedenwald (16). Pliable vessels, a high systolic blood pressure, a relatively wide pulse pressure, and reduced peripheral resistance undoubtedly are important factors which tend to produce tortuosity in coarctation. At least one of these, a high systolic blood pressure, usually becomes more pronounced with increasing duration of the disease.

Serpentine pulsation has been defined by Adler (17) as:

Pulsations in which the course of the artery shifts slightly from side to side at each heart beat, becoming manifest either as a lateral displacement of some portion of the arterial tree, or more often, in slight bending or unbending of some twist or knuckle in the artery. This type of pulsation has been called a "locomotion pulse" or "serpentine pulsation."

This type of pulsation of the arterioles was ophthalmoscopically visible in ten of the eleven patients. Aside from the frequency with which it was encountered, there appears to be nothing remarkable about the phenomenon. The factors which operate to produce tortuosity are the ones most likely to cause serpentine pulsation. Indeed, tortuosity itself may be the most important of these.

In addition to tortuosity and pulsation, varying degrees of generalized sclerosis, generalized narrowing and localized constrictions were observed in the arterioles. In our cases these changes were closely related to the severity and duration of the hypertension. A close relation is evident especially between the grade of sclerosis and the level of the diastolic pressure. These changes, in our opinion, are comparable to those encountered in the benign type of essential hypertension.

A hypertensive type of retinopathy was not encountered in any of our cases, and this observation is in accord with other reports in the literature. One patient, the oldest in the group, was found to have a partial closure of the central vein of the right eye. The blood pressure in this case was 210/116 by the indirect method, and Grade 3 sclerosis was present in the retinal arterioles.

Granström (4) has suggested that in certain cases it may be possible to differentiate between coarctation of the aorta and other types of juvenile hypertension by means of a thorough fundus examination. We feel the changes are characteristic only in the well-developed cases and that the less marked ones are suggestive but not diagnostic. Other types of hypertension seldom simulate the course of circulatory events peculiar to coarctation of the aorta.

CONCLUSIONS

Coarctation of the aorta may produce characteristic changes in the retinal arterioles.

The changes consist of spiral or "corkscrew" tortuosity and serpentine pulsation.

Varying degrees of generalized sclerosis and narrowing may develop and are closely related to the severity and duration of the hypertension.

In our experience, localized constrictions and hypertensive retinopathy are rarely encountered.

The arteriolar changes in well-developed cases may be sufficiently characteristic to aid in the diagnosis.

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DISCUSSION

DR. GORDON M. BRUCE. I should like to make two points. First, the significant finding here is serpentine pulsation. It is not necessary to stress the tortuosity, because according to the laws of dynamics serpentine pulsation cannot occur in a straight vessel. If we find this phenomenon, coexistent tortuosity goes without saying.

Second, I wish to reiterate and to emphasize Dr. Walker's implication that this finding is not pathognomonic of coarctation. Serpentine pulse is sometimes found in anemia, and not uncommonly in the normal vessels of children. Nevertheless, it furnishes a valuable diagnostic lead, and we should be grateful to the authors for bringing it so expertly to our attention.

DR. GLEN GREGORY GIBSON. This paper is an important one upon a subject important to ophthalmologists. The authors have observed and described the characteristic retinal arteriolar signs of aortic coarctation. They have concluded that, at times, these signs are sufficiently characteristic to permit a differential diagnosis between essential hypertension and this form of nonessential hypertension. Of even more importance is the fact that aortic coarctation serves as a clinical control on essential hypertension. It affords an opportunity to compare the reaction of the retinal arterioles to a simple mechanical elevation of the blood pressure, as in coarctation, with the more complex reactions of essential hypertension. These comparisons must be made with certain reservations, since this control, like so many others, has certain limitations. Among these limitations is the fact that here the diastolic blood pressure runs relatively lower than it does in essential hypertension. Another limiting factor is that there is a possibility of some renal ischemia, similar to that which occurs in the Goldblatt dog due to narrowing of the renal artery. Nevertheless, the possibility exists that the proper interpretation of the signs described by Drs. Walker and Stanfield may be helpful in arriving at a better understanding of the signs of hypertension which occur in the retina. For example, evidence is accumulating from both experimental and clinical sources that the signs of hypertensive retinal arteriolar narrowing, such as generalized narrowing and focal spastic narrowing, constitute restititional adaptations of the vascular tree to the elevated blood pressure rather than an essential component of the hypertensive sclerotic process per se. The changes in aortic coarctation tend to support this concept.

I was particularly pleased with the authors' explanation of the mechanism for the development of tortuosity. To me it clarifies much of the confusion in the literature upon the clinical significance of this sign. I would hazard a guess that tortuosity is a mechanical reaction to the elevated blood pressure, and at least initially it is unattended by the usual histological cellular alterations associated with sclerosis. This guess is based on the fact that tortuosity is a relatively common sign of coarctation and is an unimportant prognostic sign in hypertension.

DR. GLENN L. WALKER. I should like to thank the discussers for their kindness and all of you for your indulgence at this late stage in the proceedings.

I would like to answer one comment that Dr. Bruce made. It is true that we do not consider these changes as diagnostic. However, we do feel that there are very few forms of hypertension that can simulate the retinal picture of coarctation of the aorta and for this reason they may be quite characteristic.