Unusual Manifestations of Coarctation of the Aorta

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Coarctation of the aorta often is associated with other lesions or defects of the heart or great vessels which may influence the outcome of surgery. Infrequently, the coarctation occurs at an atypical site. Of the 118 cases to be reviewed in this paper, 56, or, roughly, one in two, present unusual findings. The frequency with which complicating conditions are met makes the search for additional pathology mandatory when surgical correction of the coarctation is under consideration.

Method Used in Selecting Cases

The case reports, physical findings, operative reports and postmortem findings of patients with coarctation seen at several Los Angeles hospitals were reviewed carefully. Cases in which only scanty information was available were discarded as unsuitable for the purposes of this study. There were 118 cases where the description of the coarctation and associated pathology was deemed adequate. In 56 instances, the condition was complicated by the presence of one or more additional defects.

Classic Symptoms of Coarctation

Most cases of heart failure in the acyanotic infant under 6 weeks of age are traceable to the infantile type of coarctation or aortic stenosis. The adult type of coarctation, in which the constriction is usually limited to a small segment of the aorta, is seen more commonly by the clinician. Evidences of the adult type of coarctation include: (1) reduced or absent pulsations in the abdominal aorta and femoral arteries, associated with hypertension in the upper limbs; (2) a fairly loud, rough, precordial systolic murmur of maximal intensity over the base of the heart, occurring a short but perceptible time after the first heart sound and not accompanied by a palpable thrill at the base of the heart; if the coarctation is complete the murmur may be absent; (3) collateral arterial circulation, evidenced radiographically by enlargement of the vessels in the superior mediastinum and/or upon auscultation, by bruits over the enlarged intercostal arteries and branches of the internal mammary arteries; (4) other evidence of aortic blocking, such as hypertrophy of the left ventricle and dilatation of the ascending aorta; (5) a defect or break in the continuity of the aortic outline in the left anterior oblique position, visualized either at fluoroscopy or on roentgenograms; (6) absence of the aortic knob, normally visible to the left of the spine at the level of the fifth dorsal vertebra in the adult; (7) bilateral notching (erosion) of lateral and posterior portions of the ribs; (8) abnormal asynchronism between the radial and femoral arteries: in a patient with aortic coarctation, the onset of the upstroke and the summit of the pulse wave are earlier in the radial than in the femoral artery where normally, the reverse is true; (9) a notch in the left border of the descending aorta, just at the level of the left main pulmonary artery and (10) significant diastolic vibrations reported in

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phonographic tracings from the dorsal spine of patients with coarctation of the aorta.6

**Unusual Findings on Physical Examination**

*Diminished or absent radial pulse on one side:* In coarctation of the adult type, the stenotic portion of the aorta usually lies in the vicinity of the ductus arteriosus, most often just beyond the origin of the left subclavian artery; however, coarcted areas have been found both proximal and considerably distal to this site. Absence of the left carotid and left subclavian pulses, or reduced pulsation in these vessels, suggests a coarctation situated between the origins of these vessels.

At times the coarctation is limited to the left subclavian artery alone. Such a localized coarctation may cause no symptoms whatsoever, save for an absent or diminished left radial pulse and, possibly, hypertension in the right arm. However, absence of the left radial pulse may be caused by a number of factors, such as a large cervical rib, an aortic aneurysm, or a tumor compressing the left subclavian artery.4 These and other more common conditions should be ruled out before a diagnosis of coarctation of the left subclavian artery is hazarded.

*Unilateral rib notching:* Unilateral rib notching on the right side of the body furnishes confirmatory evidence of a coarctation between the origins of the left carotid and left subclavian arteries, demonstrating that the right subclavian artery and its branches have assumed the brunt of collateral function.

If, on the other hand, the coarctation were less typically situated and had encroached upon the origin of the right subclavian artery, the left subclavian artery and its branches would be utilized to bypass the coarcted or atresic area, and this fact might be mirrored in unilateral rib notching on the left side of the body.

Infrequently, obliteration of the left subclavian artery results in unilateral rib notching.

*Pulsations below the diaphragm:* Absence of abdominal aortic pulsation and presence of the propulsion pulse in the femoral arteries suggests an atresia of the distal aorta. When the coarctation occurs below the diaphragm, it is usually located at or below the renal arteries.

**Visualization of the Coarctation**

Routine clinical methods are inadequate for the accurate localization of the level of aortic obstruction. Although, in the great majority of cases, the coarctation may be assumed to lie at or near the ductus arteriosus, the occasional occurrence of the constriction at other sites makes imperative accurate localization of the obstruction. In at least one instance, two areas of coarctation have been present in the same patient.5

Visualization of the constriction may be facilitated by means of angiocardiography or, preferably, by aortography. Occasionally, the coarcted area is visible in the left oblique position against a barium-filled esophagus. In addition to providing information concerning the degree of coarctation and the level at which the obstruction has occurred, accurate visualization of the constriction provides information concerning the length of the stenotic area and permits estimation of the distance between the orifice of the left subclavian artery and the site of the constriction; important factors in determining the feasibility of surgical correction.

**Findings in the Established Cases (118) of Coarctation**

*Coarctation as sole defect: Surgical Correction Performed:* Sixty-two patients, 46 males and 16 females, with coarctation uncomplicated by other defects and all evidencing classic symptoms of coarctation were operated upon without incident. The youngest in the group was two and one-half years old, the eldest, 49.

*Coarctation complicated by other defects: Patients who survived operation.* Eighteen patients survived operation for surgical correction of the coarctation in spite of complicating factors. In nine of these patients, the complicating condition was a patent ductus arteriosus. Descriptions of pathology found at operation may be found in table 1.

*Exploration without surgical correction:* Exploration was performed but no surgical correction was attempted in seven cases. In two
instances the coarctation was of the infantile type and was associated with a patent ductus arteriosus; in two, the coarcted segment was deemed too long for removal. In one patient aneurysms were found both proximal and distal to the coarcted area; in another, an aneurysm was discovered distal to the coarctation. One patient showed evidence of aortic and pulmonary hypertension of undetermined origin.

Operation contraindicated: Three patients were denied operation by the clinician because of excessive atherosclerosis in the aorta. Their ages were 39, 41 and 42, respectively.

Deaths: Twenty-eight of the 118 patients included in this investigation died. Six died soon after birth; an additional eight were under three months of age at the time of death. Seven patients, all over 12 months of age, died before an operation could be performed.

Three patients died during surgery? Four patients died postoperatively. An analysis of the findings at necropsy may be found in table 2.

**DISCUSSION**

As a general rule, each patient demonstrating classic signs of coarctation of the aorta should undergo resection of the coarcted area. How-
ever, the presence of additional pathology may influence outcome of the operation. A defect commonly associated with coarctation is a patent ductus arteriosus which, in the presence of severe coarctation, is in effect a compensatory mechanism similar to the enlarged intercostal arteries that make up collateral circulation. A machinery murmur, especially if accompanied by a systolic thrill, audible in the second or third intercostal space beneath the clavicle and to the left of the cardiac border, will betray the presence of a patent ductus arteriosus. Confirmatory evidence includes elevation and dilation of the main and left pulmonary arteries.

A thrill over the aortic area accompanied by a loud murmur (suggestive of associated aortic stenosis) or a diastolic murmur at the apex of the heart (indicating mitral stenosis) may escape notice: surgery of the coarctation in the presence of either of these conditions usually has a fatal outcome. Diastolic murmurs over the base of the heart should suggest the presence of an associated insufficient bicuspid valve (aortic regurgitation).

If electrocardiographic tracings show right axis deviation, the coarctation is probably associated with lesions such as aneurysm of the pulmonary artery, patent ductus arteriosus, insufficiency of the pulmonary valve, or rheumatic valvular disease with atrial fibrillation. Serious conduction disturbances may contraindicate surgery. There is a case on record, for example, of complete heart block with Adams-Stokes syndrome, occurring in an adult with bicuspid calcareous aortic valves.6 Congenital aneurysm of the cerebral vessels is frequently associated with coarctation of the aorta; therefore, the head should always be auscultated carefully.

Bacterial endocarditis and aortitis are frequently found in association with congenital bicuspid aortic valves, a frequent complication of coarctation. If the coarctation has been severe, considerable vascular damage may have occurred; or, in the older patient, vascular changes incidental to arteriosclerosis may contraindicate surgery.

CONCLUSION

Because the presence of associated abnormalities influences the outcome of surgical correction of the coarctation, intelligent pre-surgical planning must include the search for such defects. It has been the purpose of this paper to emphasize the frequent occurrence of such abnormalities, and to review those defects which most frequently accompany coarctation.

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