COARCTATION OF THE AORTA ASSOCIATED WITH A DEFECT OF THE MUSCULAR PORTION OF THE INTERVENTRICULAR SEPTUM IN AN INFANT

A Clinicopathologic Study

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THE association of coarctation of the aorta and a defect of the muscular portion of the interventricular septum has been infrequently reported. Bennetz[1] noted the case of a four-month-old infant who had coarctation of the aorta, patent ductus arteriosus, and large multiple defects of the interventricular septum. Abbott[2] in her review of 200 cases of coarctation of the aorta (adult type) found only one case associated with an interventricular septal defect located in the membranous portion.

Calodney and Carson[3] in their study of 22 cases of coarctation of the aorta in infants had necropsy reports for 21. Of those 21, 9 infants, all having coarctations of infantile type, had associated interventricular septal defects of unstated sites. Most of the nine infants also had other major defects.

We are presenting the clinicopathologic findings in an infant who had coarctation of the aorta (adult type) and a defect of the muscular portion of the interventricular septum.

CASE REPORT

Clinical Features

A 17-week-old boy was admitted to the hospital for cardiac evaluation. He had been a full-term infant weighing seven pounds. No initial respiratory distress had been noted. When he was two weeks old he was taken to the family physician because of impetigo and lack of appetite. At that time respirations were observed to be shallow; no murmur was heard.

When the infant was eight weeks old, he became colicky and respirations appeared to be labored. A roentgenogram of the chest showed a slightly enlarged heart and a large thymus. Roentgen therapy was advised and one treatment was given. After this therapy,

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the parents noted that the breathing had become fuller and more relaxed. Roentgen studies of the infant at 15 weeks showed definite cardiac enlargement.

The family history was not remarkable; two other siblings were said to be in good health.

Physical examination revealed an infant boy who was 57.4 cm. long and who weighed 5550 gm. (12.3 pounds). The rectal temperature was 101.5 degrees F. (38.5 degrees C.); the pulse was 120 per minute, and the systolic blood pressure, 140 mm. Hg. He appeared to be undernourished; the skin was gray. An abdominal type of respiration was noted with inspiratory retraction of the lower rib cage and suprasternal notch. The heart was enlarged. A grade II systolic murmur, heard best in the second and third interspaces at the left sternal border, was transmitted to the left suprascapular area. The second cardiac sound was accentuated at the left sternal border. Bounding brachial arterial pulsations were easily felt bilaterally. The liver was palpable 2 cm. below the right costal margin. No abdominal aortic pulsations could be felt. A faint femoral arterial pulsation was noted and appeared to vary with the respiratory cycle; it was present during prolonged breath-holding and during the phase of expiration. There was no clubbing of the fingers.

Laboratory findings were as follows: Hemoglobin content, 11.5 gm. per hundred milliliters; white blood cell count, 9850 per cu. mm.; fasting blood sugar, 76 mg. per hundred milliliters; and serologic tests gave negative results.

Fluoroscopic findings showed the heart to be more than 50 per cent enlarged and extending anteriorly to the chest wall, suggesting the configuration of a grossly enlarged right-ventricular outflow tract. The cardiac apex was displaced downward to the left, and in the left anterior-oblique view the posterior silhouette extended dorsally to the spine at 65 degrees of rotation. There was increased convexity of the left-ventricular silhouette with no clear-cut evidence of left auricular enlargement. A roentgenogram of the chest showed hilar markings greatly increased in size bilaterally, and the pattern of the peripheral lung fields suggested generalized increased vascularity.

An *electrocardiogram* revealed a gross delay in conduction in leads from the right precordium, with small early r waves and tremendously elevated late secondary r waves. In addition, there was a deeply inverted T wave with a delayed r wave in leads from the left precordium, a pattern suggesting the presence of both right and left ventricular enlargement.

The patient was observed for a seven-day period. On the second day in the hospital his temperature rose to 104 degrees F. (40 degrees C.) but returned to normal. The ventricular pulse rate varied from 140 to 200 beats per minute. During a severe crying spell the child's skin showed generalized mottling and moderate cyanosis. Moist râles were persistently present over both hilar areas posteriorly and in both lung bases. There was a trace of periorbital edema. The patient was treated with Mercuhydrin,* Crystodigin** and aqueous penicillin; the result was an effective diuresis and the loss of 1 kg. (2.2 pounds) in weight.

Cardiac catheterization. On the infant's fifth day in the hospital, catheterization was performed by way of the right superficial femoral artery and vein. The findings are summarized in the Table. Repeated attempts to catheterize a patent ductus arteriosus from the aorta were unsuccessful. An obstruction in the aorta below the left subclavian artery was demonstrated by retrograde aortic catheterization. Venous catheterization revealed the right auricle to be dilated. There was no demonstrable left-to-right shunt at

**Mercuhydrin, Lakeside Laboratories, Inc., 1707 E. North Ave., Milwaukee, Wisconsin.*
***Crystodigin, Eli Lilly & Co., 740 S. Alabama St., Indianapolis, Indiana.*

the auricular or ventricular levels. The blood pressure in the right auricle and in the right ventricle was abnormally high; the ventricular systolic pressure closely approximated intra-aortic pressure above the coarctation. The systolic pressure in the aorta below the coarctation was substantially lower than the right ventricular systolic pressure, ruling out the possibility of the entrance of the patent ductus arteriosus into the aorta below the coarctation.

Table.—*Findings from cardiac catheterization of right superficial femoral artery and vein*

Sampling location	Blood pressure, mm. Hg	Oxygen content	
		Volume %	% saturated
1. Inferior vena cava	—	3.7	30.8
2. Right auricle	6.2 (mean)	2.8	23.3
3. Right ventricle	118/80	2.7	22.5
4. Thoracic aorta (below left subclavian artery) above coarctation	120/68	4.3	35.8
5. Distal transverse aortic arch above coarctation	110/50	4.7	39.2
6. Left subclavian artery above coarctation	100/49	5.5	45.8
7. Thoracic aorta below coarctation	63/44	5.5	45.8
OXYGEN CAPACITY		12.0	100.0

The circulation time was 4.0 seconds (normal, 4.0 to 6.0) as measured from the right ventricle to the tongue after the injection of 2.5 ml. of fluorescein. Although the circulation time from the right ventricle to the tongue was at the lower limit of normal, it was much shorter than would be anticipated in the presence of congestive cardiac failure. The oxygen content of blood taken from the aorta above and below the coarctation was higher than that found in the right ventricle, but lower than the normal 94 to 96 per cent of capacity. This degree of oxygen unsaturation must have been due in part to a right-to-left shunt at the ventricular or proximal aortic levels and was accentuated by inadequate uptake of oxygen by the lungs in the presence of pulmonary edema.

It was concluded that the child had either an interventricular septal defect or a large patent ductus arteriosus proximal to the coarctation. Since the child's functional status was rapidly becoming incompatible with survival, surgical exploration was indicated.

Surgical procedure. A thoracotomy was performed on the seventh day in the hospital. The heart was found to be enlarged; an incision was made in the pericardium and 15 to 20 ml. of clear straw-colored fluid was removed. During mobilization of the distal aortic arch, profound bradycardia occurred. Intravenous atropine was administered, and a fast but regular pulse was established. No coarctate segment was recognized by direct visualization or by palpation. A large ductus arteriosus, in which arterial pulsations were believed to be palpable, was ligated. The pericardium was opened further to visualize the origin of the aorta. Profound bradycardia recurred, followed by cardiac arrest. More than 30 minutes of cardiac massage was required to re-establish a spontaneous heart beat. The chest was closed after a period of observing the heart beat without the aid of massage.

Twelve hours after surgery, the cardiac rhythm was regular, the ventricular pulse rate being 140 to 160 beats per minute. There were excellent brachial arterial pulsations; strong femoral pulses still were absent. Coarse rhonchi throughout the left lung, a

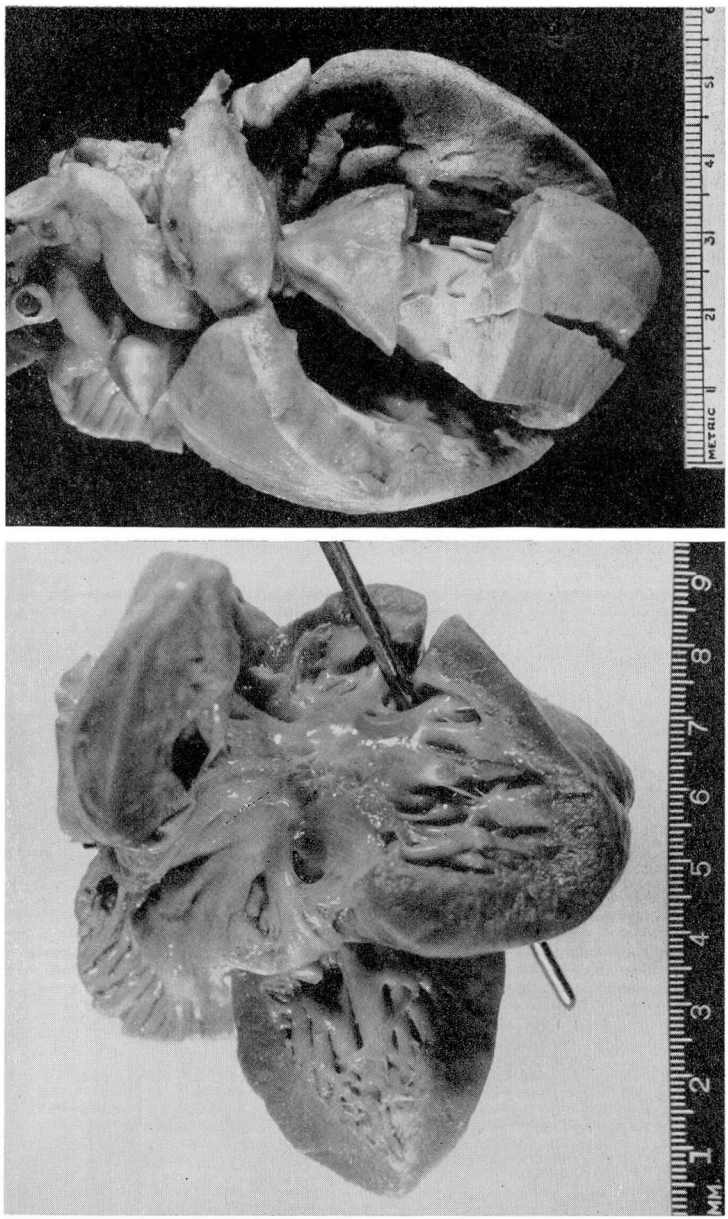


Fig. 1. Gross cardiac findings: (a) Interior view of the heart from the right side of the interventricular septum; the probe shows the location of the defect. (b) Partially opened heart; anterior window has been removed to show the prominent bilateral ventricular hypertrophy and the defect in the muscular portion of the interventricular septum.

pericardial friction rub, and a grade II to grade III rough systolic murmur at the left second to fourth interspaces were noted. There was slight periorbital edema and one plus cyanosis. Sixteen hours following surgery, the Moro embrace and the sucking reflexes were present. Twenty hours postoperatively the respirations suddenly ceased and the patient died.

Pathologic Features

The positive findings of the necropsy were limited to the heart and the lungs.

The *heart* weighed 100 gm. and was symmetrically enlarged. The myocardium was reddish brown in color and firm; the endocardium was smooth and glistening. The walls of the right and the left ventricles each measured 1.1 cm. in thickness. The cardiac valves were normal and measured circumferentially: tricuspid valve, 4.5 cm.; pulmonic valve, 3.0 cm.; mitral valve, 3.2 cm.; aortic valve, 2.0 cm. An oval interventricular septal defect, 0.9 by 0.4 cm., was present in the muscular portion of the septum, lying in the anterosuperior area 3.0 cm. from the apex (Fig. 1). The aorta was normally positioned but narrowed from a point just proximal to the origin of the left subclavian artery to immediately distal to the region of insertion of the ductus arteriosus. The ductus arteriosus was widely patent on the pulmonary arterial side but was anatomically closed at its aortic junction. At the origin of the left subclavian artery, the circumference of the aorta was 1.1 cm.; in the region of the ductus it was 0.7 cm.; and just distal to the ductus it was 1.5 cm. The opened aorta possessed a circumferential wedge-shaped area of thickening at the level of insertion of the closed ductus arteriosus, narrowing the lumen to 0.4 cm. in circumference (Fig. 2). Microscopically, sections of this area showed only

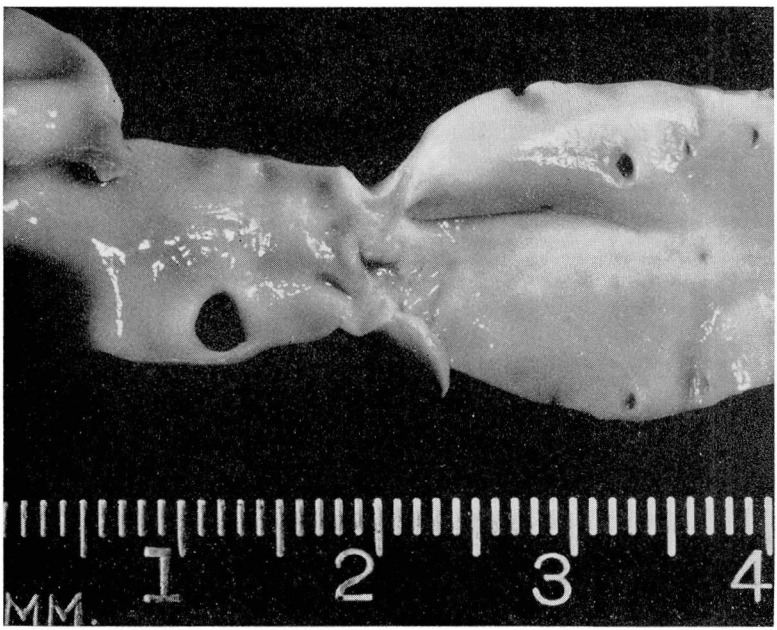
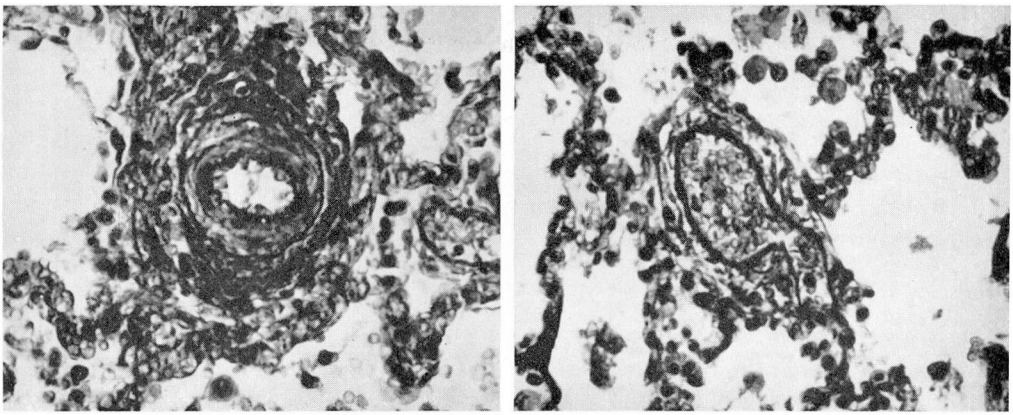


Fig. 2. Opened aorta showing the coarctate annulus of elastic tissue and slight poststenotic dilatation. The puckered insertion of the ligamentum arteriosum is centered in the defect.



(a) (b)

Fig. 3. Histologic comparison of pulmonary arterioles; photomicrographs of: (a) pulmonary arteriole showing muscular hypertrophy and some elastic fragmentation, from infant whose case is reported here; Verhoeff elastic-Van Gieson stain; X270. (b) Normal pulmonary arteriole from infant of same age; Verhoeff elastic-Van Gieson stain; X270.

thickening of the elastic media; there was no intimal change. The pulmonary artery was dilated to a circumference of 3 cm.

The left *lung* weighed 55 gm.; the right weighed 75 gm. The cut surfaces were red and somewhat firm. Tissue sections demonstrated intrapulmonary arteriolar hypertrophy characterized by thickening of the musculature of the media (Fig. 3).

The final anatomical diagnoses were: Congenital heart disease, with coarctation of the aorta (adult type), and interventricular septal defect of the muscular portion of the septum; cardiomegaly; intrapulmonary arteriolar hypertrophy.

COMMENT

Coarctation of the aorta is not commonly a clinical problem in very young persons, unless some associated difficulties are present. These associated problems include patent ductus arteriosus, intracardiac anomalies, intercurrent infection, or the development of an ectopic tachycardia. Our case was characterized clinically by stunted growth and development, and congestive failure. The infant's heart working against the resistance of the coarctate segment apparently functioned as though it were a three-chambered organ because of the interventricular septal defect. As a consequence, generalized myocardial hypertrophy and insufficiency resulted.

The apparent evidences of a right-to-left shunt, manifested by oxygen desaturation and possible shortening of the circulation time, strongly suggest shunt-reversal in the presence of myocardial failure. It is obvious that the shunt of blood originally must have been left to right and became intensified by the increased resistance, offered by the coarctation, to blood being ejected from the

left ventricle. The hypertrophic changes in the intrapulmonary arterioles probably represent a retention of fetal characteristics functioning as a protective mechanism against the pulmonary hypertension.

Intrapulmonary arteriolar hypertrophy has been reported by Edwards, Douglas, Burchell, and Christensen[4] in a case of coarctation of the aorta distal to the mouth of the patent ductus arteriosus. In a similar case, Johnson, Ferencz, Wiglesworth, and McRae[5] found that principally the small and medium-sized pulmonary arteries were involved.

The difficult problem of recognizing the coarctate segment at surgery well emphasized how subtle the external manifestations of the lesion can be in early infancy. The associated arrhythmia curtailed the surgeon's efforts to establish the true nature of the disorder.

Summary

An unusual case of coarctation of the aorta associated with an interventricular defect of the muscular septum has been presented. The presence of the latter defect is believed to be responsible for the patient's developing cardiac decompensation early in infancy.

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