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CONGENITAL HEART DISEASE IN ADULTS

(excluding interventricular septal defects).

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Clinically significant heart disease is found in no more than one in 200 autopsies on adults* and may be encountered even less frequently in clinical practice. Yet the awareness of the possibility that heart disease in adults may be congenital has come to be relatively keen in recent years. Despite this fact, a few cases are still seen in whom clinical diagnosis seems to be unduly difficult. Reemphasis of the fact that some types of congenital heart disease are compatible with a normal life span, and that such cases may mimic acquired heart disease is periodically desirable.

The most frequent congenital defect in adults is unquestionably atrial septal defects of the secundum type. Kelly and Lyons reported them to be 8 times more frequent than any other type of defect in patients aged 50 or more. Forty-three per cent of Paul Wood's 162 cases** were 21 years old or older. The adult type of aortic coarctation and patent ductus arteriosus are not found as often but still occur with sufficient frequency to require their inclusion in many differential diagnostic lists. Thirty-two per cent of Wood's collection of 115 cases of patent ductus were aged 21 or more. Of the three types, coarctation is the least frequent in adults 21 or more years old but it has been reported in elderly patients.

Diagnoses in the following four patients were delayed for varying lengths of time owing in part to general unwillingness to accept diagnoses ordinarily reserved for children or young adults. In one of them, correct diagnosis before death was not achieved although the congenital nature of the lesion was accepted shortly before exitus.

* Gelfman, Raymond, and Levine, Samuel A.: The incidence of acute and subacute bacterial endocarditis in congenital heart disease. Amer. J. Med. Sci. 204:324-333, September, 1942.

** Wood, Paul: Diseases of the heart and circulation. Second edition. Philadelphia, Lippincott, 1956. Page 360.

Case One. [redacted], [redacted]

A 55-year-old [redacted] woman was admitted on [redacted]-62 because of intractable congestive failure. At age 6, she had had a bout of rheumatic fever but recovered fully and led a normal life until she was 27 when severe hypertension was discovered. She remained asymptomatic, however, until she reached age 42 when, while under emotional stress, she developed severe anterior chest pain, nausea and vomiting. The attacks recurred occasionally, responding indifferently to nitroglycerine and requiring hospitalization in 1955 and in 1960. She was found (in 1960) to have atrial fibrillation, normal blood pressure, decreased femoral pulses, peripheral edema, and hepatomegaly. There was enlargement of both ventricles and of the left atrium by x-ray. The ECG showed left bundle branch block. She responded fairly well to standard therapeutic measures.

She remained completely incapacitated and only barely compensated until January, 1962, when an attack of lobar pneumonia seemed to have initiated a period of steady deterioration. She was finally admitted when home care became impossible.

On admission, she had atrial fibrillation with a ventricular rate of 90; blood pressure was 150/70. She was emaciated and mildly cyanotic. Neck veins were moderately distended. There was marked cardiomegaly and a prominent precordial heave. At the apex there were a grade II blowing holosystolic murmur and a short mid-diastolic rumble. There was also a grade I decrescendo diastolic murmur along the left sternal border. The pulmonic second sound was louder than the aortic. Arterial pulses could easily be felt along the vertebral borders of the scapulae but femoral pulses were absent.

Venous pressure was 30 cm. of saline and circulation time was 35 seconds. ECG showed atrial fibrillation and left bundle branch block. Cardiac films showed enlargement of left ventricle, normal left atrium, annular calcification in the region of the mitral valve, calcification in the region of the ductus arteriosus, and a small aortic knob.

Cardiac catheterization showed moderate pulmonary hypertension, elevation of right atrial pressure, and low cardiac output.

On the 13th hospital day, the patient suddenly developed ventricular fibrillation and expired. At autopsy, the heart weighed 600 grams and showed marked left ventricular hypertrophy. The coronary vessels and cardiac valves were normal although the calcification of the mitral annulus, noted during life by the roentgenologist, was verified. The most striking feature was the presence of an adult-type aortic coarctation the lumen of which measured one or 2 mm. in

diameter. It was occluded proximally by a large thrombus. Apart from these features, the findings were those of chronic congestive failure. There was no mention of bicuspid aortic valve or of arterial anastomotic channels.

Comment

Diagnosis was delayed, not only because of the patient's age but also because of her sex. Aortic coarctation is definitely more frequent in men than in women by a ratio of 2:1 or more (Frick, et al; Reifenstein, et al). In addition, there was a reasonable likelihood that the patient had had rheumatic fever as a child and the report that severe hypertension had been discovered at age 27 further complicated the clinical picture. It would be interesting to know more precisely when hypertension made its appearance. In general, hypertension in the upper extremities (and notching of the ribs) does not develop in patients with aortic coarctation until the patient is in his teens. Thereafter it rises progressively but tends to level off in no more than a decade (Campbell and Baylis). The presence of the murmur of aortic regurgitation was used by some observers to support the diagnosis of rheumatic heart disease despite the fact that about one-fourth of all patients with aortic coarctation have some degree of aortic regurgitation (Campbell and Baylis); forty-two per cent were found by Reifenstein and colleagues to have bicuspid aortic valves at autopsy. Finally, the fact that the patient rather suddenly began to deteriorate a few months before admission, that she died in congestive failure, and that no pulses were found in the lower extremities were used by some observers, along with the fact that she had atrial fibrillation, to support the notion that she had sustained multiple arterial emboli of major magnitude. Aortic coarctation was denied despite the obvious presence of collateral channels about the scapulae.

Actually, the terminal course was fairly characteristic. Exitus was due to congestive failure in 18 per cent of the cases reported by Reifenstein and co-workers, 23 per cent being due to rupture of the aorta and 22 per cent to endocarditis. Intracranial bleeding was listed as the cause of death in 11 per cent.

The source of the embolus which terminally occluded the coarcted segment is unknown.

While definitive surgical management at the last admission was not feasible, even if the correct diagnosis had been generally accepted, it almost certainly would have been attempted had the diagnosis been made before the patient's precipitous decline began a few months before her death. Had the same findings been present in a young man there would probably have been very little delay in making the correct diagnosis. In any case, and no matter how old the patient, it is difficult to counter the clinical truism that absence of femoral pulses, plus large arterial pulsations along the medial margins of the scapulae, plus hypertension in the upper extremities means aortic coarctation.

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Case Two. [redacted]

A 60-year-old [redacted] man was admitted to the hospital for the fourth time in [redacted], 1954, complaining of severe shortness of breath and anasarca.

Review of the record disclosed that at age 15 a physician told the patient his heart was abnormal. No symptoms appeared, however, until he was about 34, at which time he developed dyspnea on exertion, ankle edema, and abdominal swelling. Treatment with digitalis and low salt diet resulted in complete recovery and he was able to resume his work which involved moderate physical exertion. There were occasional exacerbations of dyspnea and abdominal swelling but they were not incapacitating until he reached about 45 years of age, when abdominal swelling became very severe. Although treatment with digitalis and diuretics produced some improvement, the patient was never again able to work regularly. At age 54 abdominal and ankle swelling, dyspnea, and palpitation became so severe that hospitalization was sought [redacted]-49). At this time mention was made of mild substernal burning, especially after eating.

Physical examination disclosed a somewhat emaciated man who could lie flat in bed without difficulty. There was moderate distention of the neck veins in the seated position and questionable cyanosis of the nail beds. There was flatness to percussion over the base of

the right lung posteriorly. The heart was enlarged to the mid-axillary line and the cardiac impulse was diffuse, forceful and heaving. There was a diastolic thrill along the left sternal margin. The rate was 58 and the rhythm basically irregular. There was a definite paradoxical pulse. A loud, blowing diastolic murmur was audible along the left sternal margin and an equally loud rough systolic murmur was heard all over the precordium, its maximal intensity being at the apex. In the same area, experienced observers reported a short mid-diastolic rumble. P_2 was markedly accentuated. The blood pressure was 150/60 in both arms and was slightly higher in the legs. There was a definite fluid wave in the abdomen. The liver extended 6 cm. below the costal margin and was thought to be pulsate. It was not tender. The spleen was not felt. The prostate was diffusely enlarged and nodular. There was moderate edema of the ankles.

X-rays of the chest showed cardiomegaly mainly of the left ventricular type. The atria did not seem to share in the enlargement. The most striking feature of the examination was extreme dilatation of the aorta and pulmonary artery, both of which were partly calcified. There was fullness of the hilar vessels but their movements were thought to be normal. The electrocardiogram showed auricular fibrillation and occasional bouts of bigeminy. The mean QRS axis in the frontal plane was about +180 degrees, there being some evidence also of right ventricular hypertrophy.

The venous pressure was 282 mm. of saline, the decholin circulation time 62 seconds, and the vital capacity 2400 cc. The hemoglobin was 16.2 grams and red and white cell counts were normal. The total serum proteins were 5.8 gm., the A/G ratio being 3.2/2.6. BSP test showed 17 per cent retention in 45 minutes. The icteric index was slightly elevated. A Kline test was negative. Urinalyses were not remarkable.

The prevailing diagnostic impression was rheumatic heart disease with involvement of mitral, aortic, and tricuspid valves. Under vigorous diuretic measures and treatment with digitalis, the patient's weight declined from 127 lbs. (on entry) to 115 lbs. (2 weeks later) and there was marked symptomatic improvement.

For the succeeding five years, he was followed in the cardiac clinic, his main complaint being ascites. In addition to digitalis and mercurial diuretics, he received 151 abdominal taps over this period of time. Two to five liters of sterile transudate were removed each time. The murmurs changed very little. The systolic and diastolic murmurs were never continuous and the previously reported apical

diastolic rumble was reported only twice. Blood pressure readings varied from 105/48 to 140/60. There was a mild tendency to hypoproteinemia during the interval. In the last year before the terminal events, anterior chest pain, relieved by nitroglycerine, was noted.

At age 59, he was readmitted for recompensation and further study. Physical findings, x-rays, and electrocardiograms were largely unchanged. The BUN was 24 mg., serum chlorides 86.4 mEq, and serum CO_2 20.4 mEq. Serum bilirubin 0.4 mg., and cephalin flocculation test was 3+ in 48 hours. PSP test was normal. Cardiac catheterization (July, 1954) disclosed the following:

	Pressure	O_2 Content	O_2 Sat.
SVC	-	6.9	42.3 %
RA	32/14	7.1	43.7
RV	110/15	9.3	56.9
PA	110/52	11.6	71.7
Brachial art.	108/50	15.8	97.5
Capacity	-	16.2	100.0

The patient's decompensation never yielded fully to treatment. He was discharged after cardiac catheterization but had to be readmitted a few weeks later because of reaccumulation of fluid and dyspnea on exertion. Physical findings and laboratory data remained virtually unchanged. He responded fairly well to mercurial diuretics, Southey tubes, and salt-free diet. He was again discharged, only to return for the last time about six weeks later because of severe cardiac decompensation. He responded poorly to treatment and expired suddenly on the 16th day in the hospital ([redacted]-54).

At autopsy the heart weighed 1010 grams and was markedly dilated. The coronary arteries were patent throughout. The valves were not significantly abnormal. Both ventricles were markedly hypertrophied. Both aorta and pulmonary artery were dilated, the latter more than the former. A window-like opening connected the two vessels, opening into the aorta just distal to the origin of the left subclavian artery. There was no tubular ductus arteriosus as such, the opening being about a centimeter in diameter. Its margins were markedly sclerosed and calcified.

The liver showed severe portal cirrhosis and pericapsular fibrosis.

Comment

The prevailing view, when the patient was seen in 1949, was that he had rheumatic heart disease with both aortic and mitral valvular involvement. The ascites was explained on the basis of tricuspid disease and the markedly dilated pulmonary artery was largey ignored although one observer briefly considered Lutembacher's syndrome. No attempt to review the case from a fresh point of view was made until 1954, despite the clear right axis deviation in the ECG and the tremendous dilatation of the main pulmonary artery. Cardiac catheterization, undertaken 5 years after the patient was first seen, showed severe pulmonary hypertension and a progressive rise in oxygen content of blood taken from right atrium, right ventricle, and pulmonary artery. This established the lesion as congenital but failed to establish the site of the left-to-right shunt. Under such circumstances, as pointed out by Bowers and co-workers, the shunt "... may be present above, below, or both above and below the pulmonary valve." The concensus at the time was that the patient had interventricular septal defect with pulmonary hypertension and congenital aortic insufficiency.

It is not clear why congenital heart disease was not seriously entertained during the first five years of observation, nor is it clear why, after catheterization data were available, atypical ductus arteriosus received no consideration. Atypical ductus arteriosus has been recognized as a clinical entity at least since 1944 when Chapman and Robbins recognized the probable association between large left-to-right shunt of long standing through a patent ductus and the development of severe pulmonary vascular sclerosis. Their case was similar clinically to the present case except that cardiac catheterization was not available at the time. Since then, the natural history of untreated patent ductus has been more fully studied and the marked alteration in the clinical picture that occurs when pulmonary hypertension appears is well recognized (Cosh; Dailey, et al., Hultgren; Whitaker, et al.). Experience has shown clearly that ligation of a ductus arteriosus which is accompanied by significant pulmonary hypertension is very hazardous.

At autopsy, the ductus was a window type instead of the usual tubular variety. Even so, it should not be confused with aortic - pulmonary septal defects which are more akin to the truncus arteriosus deformity than to patent ductus arteriosus. The window was in the ductus position which is more distal than the aortic-pulmonary septal defect. The two anomalies can best be differentiated during life by retrograde aortography (Gasul, et al.).

Case Three. [redacted]

A 40-year-old [redacted] was admitted on [redacted]-61 for study. She had known of the presence of a heart murmur since birth but, although smaller than her nine siblings, had no disability. She noted cardiac irregularity in her teens but never required treatment. She delivered a full-term child in 1950 and in 1955 a second pregnancy was terminated by Caesarian section because of her heart. There were no signs of failure at the time, but in 1959 she noted abdominal fullness and right upper quadrant tenderness. In 1960, marked abdominal swelling, dyspnea on exertion, and orthopnea appeared and treatment for cardiac decompensation was instituted. She responded quite well and was asymptomatic when admitted for study.

On admission, the blood pressure was 105/20 and the pulse was 68 and irregular. The heart was enlarged; the pulmonic second sound was louder than the aortic and there was a nearly continuous murmur, loudest in the third left interspace, which showed marked systolic accentuation. There was mild right upper quadrant tenderness but no ther significant findings. Electrocardiogram showed atrial fibrillation and left ventricular hypertrophy. Films showed enlargement of the pulmonary artery and left atrium as well as marked increase in pulmonary vascular volume.

Cardiac catheterization showed pulmonary arterial pressure of 44/30 and a progressive increase in oxygen content of blood taken from right atrium (12.6 vpc), right ventricle (15.1 vpc) and pulmonary artery (16.9 vpc). Retrograde cineangioaortography showed a widely patent ductus arteriosus about 1.5 cm. long which opacified quite readily from the aortic end. Left atrium and left ventricle were both enlarged; both were opacified by contrast medium that passed from aorta through the ductus to pulmonary artery and thence via the pulmonary bed to the left heart.

At operation, the ductus was found and ligated with little difficulty. Chest films taken a few months after operation showed diminution in cardiac size and in the diameter of the pulmonary artery.

Comment

The second patient with patent ductus arteriosus differs from the first in that the signs were classical and there was at the time of surgery only a very slight degree of pulmonary hypertension. With

surgery, there is every expectation that the patient will recover fully; without it there is the strong probability that she would have arrived sooner or later at the stage reached by the first patient when he was about 45 years old.

Her murmur was, for all practical purposes, continuous and merited the adjective machinery. It was the continuous murmur originally associated with patent ductus arteriosus by Gibson* and still called by his name in Britain. The history of mild congestive failure was not particularly unusual but suggested a somewhat ominous outlook.

The diagnostic confusion in the case probably centers on the enlargement of the left atrium which was seen on routine chest films and in cineangiograms. Yet this feature is entirely in keeping with patent ductus. Donavan and colleagues found it roentgenologically in 32 of 43 cases and Perloff and Harvey agree. Left atrial enlargement has been found to regress markedly after ligation of the ductus.

As was the case with the previous patient, cardiac catheterization failed to identify the level at which left-to-right shunting was occurring. Retrograde aortography not only demonstrated the ductus but permitted fair estimate of its size.

* Gibson, G. A.: Clinical lectures on circulatory affections. Lecture I. Persistence of the arterial duct and its diagnosis. Edinburgh Med. J., n.s. 8:1-10, 1900.

Case Four. [redacted] [redacted]

A 56-year-old [redacted] man was admitted on [redacted]-61 complaining of dyspnea on exertion. He had known of the presence of some type of heart disease since he was 19 years old, and had had dyspnea on extreme exertion since his mid-twenties. Ten years before this admission he was treated in a South Carolina hospital for heart failure and rapid pulse rate. He recovered and was able to work as a salesman until 1960 when fatiguability and dyspnea became extreme. He was admitted to [redacted] three times during 1960 because of accumulation of fluid. The admission to [redacted] in [redacted], 1961, was for detailed study.

Physical examination disclosed moderate emaciation but no obvious distress. The heart was moderately enlarged and there was an irregular rhythm. Blood pressure was 110/70. The pulmonic second sound was accentuated and split. There was a grade III rough systolic murmur maximal along the left sternal border but also easily heard at the apex. The liver was slightly enlarged but there was no edema.

Venous pressure and circulation time were both at the upper limit of normal. The elctrocardiogram showed right bundle branch block and atrial fibrillation. Chest films showed cardiomegaly and signs of marked increase in pulmonary vascular volume. The pulmonary artery was tremendously dilated and contained calcific deposits. Both ventricles seemed to be enlarged. Cardiac catheterization showed a marked increase in oxygenation of blood taken from right atrium as compared with that from the superior vena cava (13.6 and 8.7 volumes per cent, respectively). The pulmonary artery pressure was 70/45, pulmonary flow was 6.9 l./min. The right atrial mean pressure was 9 mm Hg.

Diagnosis was interatrial septal defect, secundum type, and mild pulmonary hypertension. Surgical intervention was thought to be unwise because of the presence of pulmonary hypertension and the patient's age. The patient was discharged a month after admission. Two weeks later ([redacted]-62) he was seen in the emergency room complaining of pleuritic chest pain but was not readmitted. Death came suddenly on [redacted]62, over 10 years after the appearance of congestive failure and about 2 years after the patient became totally disabled.

At autopsy, the heart weighed 720 grams. Both ventricles showed hypertrophy, the right more than the left. There was a secundum defect between the atria measuring nearly 2 cm. in diameter. The pulmonary artery was massively dilated and showed severe sclerosis in all its major branches. There were numerous old and recent emboli in the smaller radicles of the pulmonary tree.

Comment

The case is typical of the fate awaiting most patients who go through life with an uncorrected sizeable defect of the atrial septum. Although the lesion is compatible on rare occasions with normal life span (several patients are known to have lived to be 80 or more) it much more commonly produces pulmonary hypertension in the forties or fifties. The report by Campbell et al., shows that if patients with atrial septal defects survive the first 2 years of life, they are likely to be asymptomatic at least to age 20. But by age 50, less than a fourth of such patients remain asymptomatic [and most are in serious trouble]. These comments apply mostly to the secundum type defects, although Bedford reports 2 patients with primum defects who were over 60.

The clinical diagnosis of atrial septal defect and the differentiation between primum and secundum types during life have now reached a high degree of refinement. In adults, the development of dyspnea on exertion (and atypical chest pain *), together with basal systolic murmur, accentuation and splitting of the pulmonic second sound, and complete or incomplete right bundle branch block, are highly suggestive of atrial septal defect of the secundum type. Barber and colleagues lay special emphasis on wide splitting of the second sound in such cases. Dimond and Benchimol describe the basal systolic murmur as beginning immediately after the first sound and often ending before the second. These, it should be emphasized, are impressions gained primarily from phonocardiograms. For those who still find it necessary to use the stethoscope, it suffices to note that the basal systolic murmur of atrial septal defect, secundum type, does not obscure the first or second sound, the latter being characteristically accentuated. When pulmonary flow is relatively low and the pulmonary artery not grossly dilated, the systolic murmur is usually blowing; when flow is high, it often becomes raucous and rough. The electrocardiogram (Dreifus, et al.) is almost never normal; intraventricular conduction defects and various versions of right-sided patterns are the rule, so much so that their absence casts doubt on the diagnosis.

Primum defects (common AV canal deformity) are associated with different murmurs and with QRS loops (frontal) that rotate counterclockwise rather than the reverse as is the case with secundum defects.

The natural history of secundum defects is primarily determined by the size and duration of the left-to-right shunt precisely as seems to be the case with patent ductus arteriosus and with uncomplicated ventricular septal defects. The end stage for all of them is pul-

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* Chapman, C.B., and Fraser, R.: Clinical and hemodynamic features of uncomplicated interatrial septal defect in adults. Amer. Heart J. 46:352-363, September, 1953.

monary vascular sclerosis and hypertension, partial reversal of the shunt, and congestive failure. Bacterial endocarditis may supervene in cases of patent ductus and ventricular septal defect but is less likely in cases of atrial septal defect. It does occur, however, and a recent report on the point is available (Griffiths).

Repair of secundum defects has become feasible and should be undertaken before significant pulmonary hypertension develops.

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Conclusions

Congenital heart disease, although relatively rare in adults, is important in that it is often still correctible. As a matter of general principle, signs and symptoms often become less characteristic with the passage of years but few types of congenital heart disease completely lose their identity in adults. Had diagnosis been made earlier in the patient with aortic coarctation - and there seems to be no adequate reason for such a long delay - correction almost certainly would have been possible.

In the case of large left-to-right shunts the final common pathway is pulmonary sclerosis and hypertension leading to congestive failure and death. This dire sequence was undoubtedly prevented in one of the cases of patent ductus arteriosus in which diagnosis was made and surgery done before irreversible changes took place. In the other case of patent ductus, and in the case of atrial septal defect, the disorders ran their natural, untreated, courses with congestive failure and exitus in the sixth decade as the end-result.

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Atrial septal defect: 225 cases, 22 deaths
Coarctation of aorta: 173 cases, 17 deaths
Patent ductus: 620 cases, 21 deaths

Bowers, D., Burchell, H. P., and Wood, E. H.: Difficulty in the precise localization by cardiac catheterization of left-to-right shunts near the pulmonary valve. Proc. Staff Meetings, Mayo Clinic 30:261-266, June 15, 1955.

"When, in the same patient, the oxygen content of the blooc in the pulmonary artery is higher than that in the right ventricle, and the oxygen content of the blood in the right ventricle is itself higher than that in the right atrium, left-to-right shunts may be present above, below, or both above and below the pulmonary valve." Three well-studied cases included.

Kjellberg, S. R., Mannheimer, E., Ruhde, V., and Jonsson, B.: Diagnosis of Congenital Heart Disease. Chicago, Year Book Publishers, 1955.

Onat, T.: Die Volumenbelastung des rechten Ventrikels. Röntgenologische, phonokardiographische und elektrokardiographische Korrelationsstudie. Cardiologia 39:191-218, September, 1961.

Study based on 44 cases of atrial septal defect (some with anomalous pulmonary venous connection). Contains a very useful correlation of frontal and horizontal QRS vectors, and right ventricular output.

B. Aortic Coarctation

Bertin, R. J.: Treatise on the Diseases of the Heart and Great Vessels. Edited by J. Bouillaud. Translated by C. W. Chauncey. Philadelphia, Carey, Lea, and Blanchard, 1833.

Author quotes several cases of aortic coarctation, the oldest 57, from other writers.

Bramwell, C., and Jones, A. Morgan: Coarctation of the aorta: the collateral circulation. Brit. Heart J. 3:205-227, 1941.

Bunnell, I. L., Ikkos, D., Rudhe, U. G., and Swan, H. J.: Left-heart volumes in coarctation of the aorta. Amer. Heart J. 61:165-172, February, 1961.

Authors found highly efficient LV ejection patterns which were not affected by operation.

Campbell, M., and Baylis, J. H.: The course and prognosis of coarctation of the aorta. Brit. Heart J. 18:475-495, October, 1956.

One hundred thirty cases, 80 followed for at least 5 years. One-fourth had aortic regurgitation. Hypertension develops during early teens and levels off at 18 years or so. Sudden death, usually in the third decade, occurred in 6 cases.

Campbell, M., and Polani, P. E.: The aetiology of coarctation of the aorta. Lancet 1:463-468, March 4, 1961.

Inconclusive study of family backgrounds of 151 patients.

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The ratio of men to women is probably about 2 to 1. Children may have no notching of the ribs. Blood pressure begins to rise quite early and the greatest increase occurs between the ages of 10 and 20. After age 23, there was very little change in the group studied.

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About 150 patients with co-arctation and pregnancy have been reported. In the group, there were 380 pregnancies. Mortality was about 3.5 per 100 pregnancies. Rupture of the aorta was the most common cause but the author does not believe the usual hazards of co-arctation are increased by pregnancy.

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Gough, J. H.: Coarctation of the aorta in father and son. Brit. J. Radiol. 34:670-672, October, 1961.

Gudbjerg, C. E., and Petersen, O.: Coarctation of the aorta. Relation between roentgenologic and hemodynamic findings. Radiology 75:399-405, September, 1960.

Males predominate. Notching is rarely seen until 11 to 14 years; over 14, it is almost the rule. The higher the syst. gradient, the greater the notching.

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Coarctation of the aorta (Albrecht Mickel, 1827). Idem 9:307-311, February, 1962.

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Coarctation of the aorta (Legrand, 1833). Idem 10:266-271, August, 1962.

Kenner, Thomas: Der arterielle Puls bei Aortenisthmusstenose. Z. Kreislaufforsch. 48:730-734, August, 1959.

Usual features of the arterial pulse wave distal to coarctation:

(a) Practically triangular.
(b) Straight descending limb.
(c) Absent dicrotic notch.

Using a model of tubes, the author reproduced the wave as seen clinically. With increasing stenosis in the middle of the tube, the relative content of higher harmonics in the central and peripheral pulse decreases. By this means the pulses become like those in the Windkessel. Pulse wave velocity is not altered.

March, Harold W., Hultgren, Hubert N., and Gerbode, Frank: Immediate and remote effects of resection of the hypertension in coarctation of the aorta. Brit. Heart J. 22:361-373, June, 1960.

Eighty patients operated at Stanford, 58 with complete records. At discharge, 33 per cent had lost their hypertension. In the others (unless there was aortic regurgitation or residual coarctation) the blood pressure ultimately fell to normal.

The oldest patient was 55.

Oey, F. T., and Noordijk, J. A.: Coarctation of the aorta in older patients. Thorax 16:169-175, June, 1961.

The average age of untreated patients is about 35 years.

The authors studied 32 patients who were operated after age 35. Two died at surgery. A graft was used in only one. Results in the other 30 were very good. The oldest survivor was 49.

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Includes original clinical observations on coarctation in adults. Peacock's is the first textbook in English on congenital heart disease.

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The probable incidence of the adult type is 1:3000 autopsies. Oldest patient in the series was 76. The ratio males:females was 5:1. Seventy-four per cent died of rupture of the aorta, bacterial endocarditis, congestive failure, or intracranial hemorrhage. The aorta may rupture proximal (19 cases) or distal (5 cases) to the coarctation. Bicuspid aortic valve was present in 42 per cent.

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The patient had known she had heart disease all her life. She died following surgery for incarcerated femoral hernia.

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Baronofsky, I. D., Gordon, A. J., Grishman, A., Steinfeld, L., and Kreel, I.: Aorticopulmonary septal defect; diagnosis and report of case successfully treated. Amer. J. Cardiol. 5:273-276, February, 1960.

Bonham Carter, R. E., and Walker, C. H. M.: Continuous murmurs without patent ductus. Lancet (London) 268:272-276, February 5, 1955.

The authors present five cases of children with continuous murmurs attributable to ventricular septal defect (4 cases), and aortic coarctation with large collaterals (one case). Cardiac catheterization showed O_2 content of blood from PA to be significantly higher than that in RV blood in 2 cases. They list the following causes of continuous murmurs at the base of heart: (1) patent ductus; (2) aortic pulmonary window; (3) pulmonary atresia or truncus arteriosus, where lungs derive blood supply from aorta; (4) continuous venous hum; (5) arterio-venous aneurysms; (6) communication of sinus of Valsalva with PA; (7) IV septal defect, usually with associated deformity of aortic valve.

Bouillaud, J.: Traité clinique des maladies du coeur, précédé de recherches nouvelles sur l' anatomie et la physiologie de cet organe. Paris, J. B. Baillière, 1835. Vol. 2, p. 562.

Describes a woman who died at 28 from "atypical" ductus arteriosus. Also presents several cases of atrial septal defect but none in adults.

Burman, D.: Familial patent ductus arteriosus. Brit. Heart J. 23:603-604, September, 1961.

Patient's father and 2 sisters had patent ductus arteriosus.

Chapman, C. B. and Robbins, S.L.: Patent ductus arteriosus with pulmonary vascular sclerosis and cyanosis. Ann. Int. Med. 21:312-323, August, 1944.

Cosh, J. A.: Patent ductus arteriosus with pulmonary hypertension. Brit. Heart J. 15:423-429, October, 1953.

Cotton, A. C.: Report of a case of anuria. Arch. Pediat. 16:774-778, October, 1899.

First case of aorticopulmonary septal defect in the American literature. The patient was a newborn who died 5 days after birth. The case as described was actually one of truncus arteriosus, in which aorta and pulmonary artery formed a common trunk.

Dailey, F. H., Genovese, P. D., and Behnke, R. H.: Patent ductus arteriosus with reversal of flow in adults. Ann. Int. Med. 56:865-882, June, 1962.

Donovan, M. S., Neuhauser, E. B. D., and Sosman, M.: Roentgen signs of patent ductus arteriosus. Amer. J. Roentgenol. 50:293-305, September, 1943.

"A dilated left auricle is one of the commonest signs of a patent doctus arteriosus ..." It was present in 32 of 43 cases.

Downing, D. F., Bailey, C. P., Maniglia, R., and Goldberg, H.: Defect of the aortic septum. Amer. Heart J. 45:305-314, February, 1953.

Fishman, L.: Patent ductus arteriosus in a patient surviving to seventy-four years. Amer. J. Cardiol. 6:685-688, September, 1960.

Gasul, B. M., Fell, E. H., and Casas, R.: The diagnosis of aortic septal defect by retrograde aortography: Report of a case. Circulation 4:251-254, August, 1951.

The authors hold that retrograde aortography is the only method of differentiating aortic septal defect from patent ductus arteriosus.

Gintrac, E.: Observations et recherches sur la cyanose ou maladie bleue. Paris, J. Pinard, 1824.

Gross, R. E.: Surgical closure of an aortic septal defect. Circulation 5:858-863, June, 1952.

First successfully treated case of the lesion. The patient, a girl aged 4 at the time of surgery, was well 3 years afterward.

Hackney, J. L.: Persistent patent ductus arteriosus in an adult. A case report. J. Oklahoma Med. Asso. 55:327-329, August, 1962.

Hubbard, T. F., and Neis, D. D.: The sounds at the base of the heart in cases of patent ductus arteriosus. Amer. Heart J. 59:807-815, June, 1960.

Hultgren, H., and four co-authors: The syndrome of patent ductus arteriosus with pulmonary hypertension. Circulation 8:15-35, July, 1953.

Keith, T. R., and Sagarminaga, J.: Spontaneously disappearing murmur of patent ductus arteriosus. A case report. Circulation 24:1235-1238, November, 1961.

Krovetz, L. J., and Warden, H. E.: Patent ductus arteriosus. An analysis of 515 surgically proved cases. Dis. Chest 42:46-57, July, 1962.

McCutcheon, F. B., Hara, M., Lincoln, B. M., and Dungan, W. T.: Patent ductus arteriosus. An analysis of eighty cases. J. Arkansas Med. Soc. 59:97-102, August, 1912.

Neufeld, H. N., Lester, R. G., Adams, P., Jr., Anderson, R. C., Lillehei, C. W., and Edwards, J. E.: Aorticopulmonary septal defect. Amer. J. Cardiol. 9:12-25, January, 1962.

Authors report 6 cases of their own and report 60 others from the literature. The difficulty of distinguishing between ventricular septal defect, aorticopulmonary septal defect, and patent ductus is emphasized. Selective aortography is the most satisfactory method.

Three of 5 patients who were operated survived.

Perloff, J. K., and Harvey, W. P.: Unusual left atrial enlargement with patent ductus arteriosus. Amer. Heart J. 60:804-810, November, 1960.

Patient was a 37-year-old woman with classical ductus arteriosus. Pulmonary flow was 15 liters (5 times the systemic). Left atrium was markedly enlarged by x-ray and regressed after operation.

Silver, A. W., Kirklin, J. W., Ellis, H. E., Jr., and Wood, E.: Regression of pulmonary hypertension after closure of patent ductus arteriosus. Proc. Staff Meetings Mayo Clin. 29:293-300, May 19, 1954.

Shows reversibility in 4 cases of pulmonary hypertension as result of ligation of ductus.

Somerville, J.: Aortopulmonary septal defect; five cases treated by operation. Guys Hosp. Rep. 108:177-193, 1959.

Aortic pulmonary window always lies between ascending aorta and the pulmonary trunk (not in ductus position). Related to truncus deformities.

Correct diagnosis was made in only 2 of 5 cases. Atypical ductus was the usual diagnosis (3 of 5). In only one was there a continuous murmur. X-rays were similar to those usually seen in ductus except that the aortic knob was smaller. Electrocardiograms showed left ventricular hypertrophy. Three of the 5 also showed rSR in V-one.

Spencer, H., and Dworken, H. J.: Congenital aortic septal defect with communication between aorta and pulmonary artery; case report and review of literature. Circulation 2:880-885, December, 1950.

Whitaker, W., Heath, D., and Brown, J. W.: Patent ductus arteriosus with pulmonary hypertension. Brit. Heart J. 17:121-137, April, 1955.

Best article so far. Symptoms and signs mainly those of pulmonary hypertension.

D. Atrial Septal Defect

Barber, J. M., Magidson, O., and Wood, P.: Atrial septal defect with special reference to the electrocardiogram, the pulmonary artery pressure and the second heart sound. Brit. Heart J. 12:277-292, July, 1950.

Sixty-two cases. "Wide-splitting of the second heart sound is attributed to right bundle branch block [complete or incomplete] or to delay in the emptying time of an over-filled right ventricle, and is a far more important sign of ASD than accentuation of the pulmonary second sound."

Bedford, D. Evan: Atrial septal defects. Proc. Roy. Soc. Med. 54:779-781, September, 1961.

Four hundred cases studied. Excluding those that died in infancy, about 10 per cent were primum variety. Three with primum defects were over 60 years old.

Bedford, D. Evan: The anatomical types of atrial septal defect. Their incidence and clinical diagnosis. Amer. J. Cardiol. 6:568-574, September, 1960.

Clear exposition of anatomical types, along with roentgenographic and electrocardiographic features of each.

Besterman, Edwin: Atrial septal defect with pulmonary hypertension. Brit. Heart J. 23:587-598, September, 1961.

Forty-one cases. Complete clinical analysis.

Burrett, J. B., and White, P. D.: Large interauricular septal defect with particular reference to diagnosis and longevity: report of 2 new cases. Amer. J. Med. Sci. 209:355-364, March, 1945.

Campbell, M., Neill, C., and Suzman, S.: The prognosis of atrial septal defect. Brit. Med. J. 1:1375-1383, June 15, 1957.

The authors studied 100 patients with atrial septal defect. In their view, 95 per cent (of those surviving 2 years) do well to age 20 and 85 per cent to age 30 (probably all secundum type). Less than 25 per cent are asymptomatic at age 50.

About one-tenth have anomalous pulmonary venous drainage from the right lung to atrium or veins. Another one-tenth has some degree of pulmonic stenosis.

Chiong, M. A.: Interatrial septal defect and longevity. Canadian Med. Asso. J. 83:1012-1014, November, 1960.

Case of a 74-year-old man with cyanosis, orthopnea, cardiomegaly, loud P_2, apical systolic murmur and diastolic rumble. Severe chest pain before death. Right bundle branch block and mild polycythemia were also present.

At autopsy, the heart weighed 600 grams. There was a secundum defect measuring 4.5 cm. in diameter. Pulmonary vascular sclerosis and pulmonary infarcts were also found.

Colmers, R. A.: Atrial septal defects in elderly patients: report of three patients aged 68, 72, and 78. Amer. J. Cardiol. 1:768-773, June, 1958.

Coulshed, N., and Littler, T. R.: Atrial septal defect in the aged. Brit. Med. J. 1:76-80, January 12, 1957.

Cases presented were aged 79, 68, 67, 58, and 58 years. All appear to have been secundum types with characteristic ECG changes.

Dimond, E. G., and Benchimol, A.: Phonocardiography in atrial septal defect: correlation between hemodynamics and phonocardiographic findings. Amer. Heart J. 58:343-356, September, 1959.

Thirty-two patients with secundum defects. Close splitting of second sound at the base was a main feature; it usually widened on inspiration. Systolic murmur (pulmonic) all started immediately after first sound but often ended before aortic second sound. Low-frequency diastolic murmur was heard at the apex and left sternal border in 14 patients.

Dreifus, L. S., Bender, S., Goldberg, H., and Downing, D. F.: The electrocardiogram in atrial septal defect. Dis. Chest 36:521-527, November, 1959.

Predominant V-one pattern is rsR' (secundum type). After surgery QRS does not shorten but R decreases in amplitude.

DuShane, J. W., Weidman, W. H., Brandenburg, R. O., and Kirklin, J. W.: Differentiation of interatrial communications by clinical methods: ostium secundum,

ostium primum, common atrium, and total anomalous pulmonary venous connection. Circulation 21:363-371, March, 1960.

Four forms of interatrial communication: (1) Ostium secundum, (2) Ostium primum, (3) Common Atrium, and (4) Total anomalous pulmonary venous connection.

All patients with secundum defect (74 out of 128 total) had usual murmur and splitting of the second sound at the base. All also had some degree of incomplete RBBB.

Primum defects were associated with a QRS loop (frontal) that rotated in a counterclockwise direction most of it being above the isoelectric line or describing a figure-of-eight along the line with the initial part of the loop rotating counterclockwise. Findings are similar in patients with common atrium. In patients with anomalous pulmonary venous connections (total), loops are similar to those with the secundum type defect.

Effler, D. B., and Groves, L. K.: Pitfalls in the surgical closure of atrial septal defect. Based on experience with one hundred and fifteen cases. Cleveland Clin. Quart. 28:166-175, July, 1961.

Ellis, F. H., Jr., Brandenburg, R. O., and Swan, H. J.: Defect of the atrial septum in the elderly. Report of successful surgical correction in five patients sixty years of age or older. New England J. Med. 262:219-224, February 4, 1960.

Five patients, aged 60 or more, had atrial septal defects and were submitted to surgery. All survived and were symptomatically improved.

Griffiths, S. P.: Bacterial endocarditis associated with atrial septal defect of the ostium secundum type. Amer. Heart J. 61:543-547, April, 1961.

Gross, R. E.: Atrial septal defects of the secundum type. Progr. Cardiovasc. Dis. 4:301-311, January, 1962.

Howitt, G.: Atrial septal defect in three generations. Brit. Heart J. 23:494-496, September, 1961.

Kelly, J. J., Jr., and Lyons, H. A.: Atrial septal defect in the aged. Ann. Int. Med. 48:267-283, February, 1958.

Authors reported 19 patients aged over 47 years. Atrial septal defects are eight times more frequent in patients over 50 than any other type of congenital defect.

Lewis, F. J., Winchell, P., and Bashour, F. A.: Open repair of atrial septal defects: results in sixty-three patients. J.A.M.A. 165:922-927, October 26, 1957.

Rodstein, M., Zeman, F. D., and Gerber, I. E.: Atrial septal defect in the aged. Circulation 23:665-674, May, 1961.

One of the authors' patients was 84 and one 72. Both had secundum defects (one associated with anomalous pulmonary veins) and both died in congestive failure.

Scott, R. C.: The electrocardiogram in atrial septal defects and atrioventricular cushion defects. Amer. Heart J. 62:712-714, November, 1961.

Sellors, T. H.: Atrial septal defects. Proc. Roy. Soc. Med. 54:781-783, September, 1961.

Three hundred and three cases treated surgically. Operative mortality in secundum defects was 3.3 per cent; it was over 10 per cent in the primum group.

Silver, A. W., and Kirklin, J. W.: Accuracy of diagnosis in surgical cases of atrial septal defect. AMA Arch. Surg. 80:241-243, February, 1960.

In 213 cases, only 6 errors have been made. In 3 of the 6 there was no atrial defect at all (at surgery). Two showed AV canal deformity and one showed anomalous pulmonary venous connection.

Sommer, L. S., and Voudoukis, I. J.: Atrial septal defect in older age groups with special reference to atypical clinical and electrocardiographic manifestations. Amer. J. Cardiol. 8:198-202, August, 1961.

Ten cases, which masqueraded as other types of heart disease, were presented. The patients, aged 42 to 70, usually showed various conduction defects in the ECG (one showed left bundle branch block) but one had normal conduction.

Soulié, P., Forman, J., Pequignot, H., and Vaysse, J.: Faut-il opérer les communications interauriculaires? Presse Med. 70:633-635, March 17, 1962.

Zuckerman, H. S., Zuckerman, G. H., Mammen, R. E., and Wassermil, M.: Atrial septal defect. Familial occurrence in four generations of one family. Amer. J. Cardiol. 9:515-520, April, 1962.

Eight cases of atrial septal defect in 4 generations of one family. Inheritance of the defect is best explained "on the basis of transmission of a dominant autosomal gene with incomplete penetrance."