MALFORMATIONS OF THE HEART INCLUDING TWO CASES WITH COMMON ATRIOVENTRICULAR CANAL AND SEPTUM DEFECTS AND ONE WITH DEFECT OF THE ATRIAL SEPTUM (*COR TRILOCULARE BIVENTRICULOSUM*) *

FRANCIS D. GUNN
FELLOW IN MEDICINE, NATIONAL RESEARCH COUNCIL
AND
JOHANNA M. DIECKMANN
(*From the Laboratory of the Buffalo General Hospital and the Department of Anatomy, University of Buffalo Medical School, Buffalo, N. Y.*)

I. COMMON ATRIOVENTRICULAR CANAL AND SEPTUM DEFECTS

Although absence of the membranous part of the interventricular septum is a rather common heart anomaly, absence of the entire membranous septum combined with a common atrioventricular canal and an aperture in the lower atrial septum is quite rare. It represents a definite type of cardiac malformation, six cases of which are to be found described in the literature. In addition Keith [1] states that he has seen fourteen others and cites two described by Griffith, so that altogether we have found descriptions of, or reference to, twenty-two such cases. As the literature has not been exhaustively studied, some cases may have been overlooked, especially from European literature. However, recent discussion of the subject by Mönckeberg [2] seems to preclude this possibility.

CASE I

Clinical Report: The patient was a girl, one year old, a Mongolian idiot, who died in the Children's Hospital (No. 1384) with symptoms of cardiac insufficiency. Clinically, signs of an enlarged heart, a systolic murmur and gallop rhythm, led to the diagnosis of a congenital heart lesion. The previous history and family history of the case are irrelevant. The Wassermann reaction was negative. The necropsy took place eight hours after death.

Anatomic Diagnoses: (1) Defect of the atrial septum, with numerous perforations of the same, and patent foramen ovale; defect of the upper part of the ventricular septum; persistence of a common atrioventricular ostium, with abnormal development of the atrioventricular valves.

* Received for publication June 24, 1927.

(2) Extensive hypertrophy and marked dilatation of the heart, especially of the right side, with fatty degeneration of the myocardium. (Weight 50 gm. in fixed condition; circumference at base, 14 cm.; transverse diameter, 4.6 cm.; base to apex, 6.5 cm.)

(3) Chronic passive congestion of the lungs, liver, and spleen.

(4) Partial fetal atelectasis of both lungs.

(5) Mongolian facies.

(6) Aplasia of the third phalanx of the left fifth finger.

Description of the Heart

The heart is large and generally dilated; its apex is rounded. The dilatation is more marked on the right side, especially in the atrium and auricular appendix. The left auricular appendix is unusually small and appears rudimentary.

When the heart is opened there appears a gross defect of the upper part of the interventricular septum and of the lower part of the interatrial septum, as well as an undivided atrioventricular canal, all four chambers thus communicating freely. The interventricular septum is totally defective in its membranous portion, presenting instead, in this region, an aperture of communication between the ventricles. The muscular septum, the upper margin of which is bow-shaped with its convexity downward, forms the lower boundary of the interventricular aperture. The anterior margin of the aperture curves sharply upward to the right semilunar aortic cusp. Anterior to the aperture the ventricular septum is normal and forms the posterior wall of the conus. The inferior margin of the aperture slopes in a gentle curve upward and backward to the posterior wall of the left atrium. From this region there extends into the aperture a finger-like process which will be described later. The extent of the aperture represents that area which is normally occupied by the small membranous interventricular septum, the membranous atrioventricular septum and the junctional region of the atrial and ventricular septa. The aperture measures 3 cm. in an anteroposterior direction, and 1.4 cm. from the line of attachment of the atrioventricular valves to the free margin of the septum.

Both sides of the septum show abundant trabeculae, many of which are exceedingly fine. If the spaces between the trabeculae are carefully probed with a fine, blunt instrument, at least half a

dozen points of communication between the two ventricles may be found in different parts of the septum.

The common atrioventricular canal is surrounded by valve cusps which are better developed on the anterior than on the posterior side. They arise from the aorta and from the fibrous junction between atria and ventricles and are attached below to papillary muscles and to the anterior and inferior boundaries of the interventricular aperture.

The anterior papillary muscle in the left ventricle is well developed and the posterior is absent. In the right ventricle all three papillary muscles, the large anterior, the anterior medial and the posterior medial, are well developed. The large anterior papillary muscle lies somewhat closer to the septum and has a rather more extensive attachment to it than is usual. The anterior medial papillary muscle, which consists of four or five groups of chordae, springs from the upper, anterior portion of the interventricular septum, or, dorsal wall of the conus, just anterior to the interventricular aperture. The two muscles comprising the posterior medial group do not stand away from the heart wall but resemble more closely the trabeculae carneae.

In the left ventricle a large cusp corresponding in position to the aortic cusp of the mitral valve is conspicuous. When viewed from the aortic side it is found to arise from the base of the aorta below the adjacent halves of the left and posterior semilunar cusps. It is attached to the anterior papillary muscle and to the anterior boundary of the interventricular aperture by chordae tendineae, and to the region between the right and posterior semilunar aortic cusps by a small fibrous sheet. The chordae which pass to the anterior margin of the aperture appear as if the membranous atrioventricular septum of a normal heart were shredded into chordae tendineae. The sheet of fibrous tissue which occupies the region between the right and posterior aortic cusps corresponds to the extreme upper portion of the normal, membranous, interventricular septum.

Laterally, the aortic cusp of the mitral is continuous with a small cusp which arises from the annulus fibrosus and which is attached to the anterior papillary muscle. Medially the large aortic cusp becomes continuous, by means of a strand of valve tissue, with a cusp in the right ventricle comparable in position to the anterior leaflet of the tricuspid. The strand is similar to those often seen

connecting cusps in the normal heart. It is attached to the upper part of the interventricular septum anterior to the aperture by one of the several groups of chordae which comprise the anterior medial papillary muscle.

The anterior and lateral cusps of the tricuspid are attached as follows: the anterior, to the anterior medial papillary muscle and to the large anterior muscle; the lateral, which corresponds to the posterior cusp of the normal heart, to the large anterior and the posterior medial muscles. This is presumably normal. The lateral cusp, however, is poorly developed in that portion which lies against the posterior wall of the ventricle, being closely applied to the wall like the papillary muscle to which it is attached.

Lying against, and closely applied to the posterior half of the interventricular septum in the right ventricle, there is a cusp corresponding in size and position to the posterior half of the medial leaflet of the tricuspid. It arises from the upper posterior portion of the interventricular septum, and is attached in two places. The attachment to the posterior papillary muscle consists of only one or two chordae; the attachment to the septum is by several chordae lying beneath the cusp and by two long chordae stretching obliquely forward to be attached to the anterior half of the septum.

There remains, finally, a rudimentary cusp, the finger-like process referred to above. It is a fibrous mass, measuring 7 mm. in length and about 3 mm. in diameter. It projects freely into the aperture and is continuous over the posterior margin of the ventricular septum with the medial leaflet of the tricuspid. From its under surface there arise chordae which are attached, not directly to the septum, but to a narrow membrane, 7 mm. long and 3 mm. high, which in turn is attached to the upper margin of the interventricular septum in its posterior half.

The atrial septum shows conspicuous malformations. It consists almost entirely of a sheet of fibrous tissue, corresponding to the valvula foraminis ovalis of the normal heart. It is perforated all around its periphery by numerous foramina ranging in diameter from 1 to 5 mm. The muscular portion of the atrial septum (limbus) is limited to the upper anterior region of the septum and passes downward to become continuous with the musculature of the anterior wall of the atrium. The limbus does not quite overlap the upper anterior curved margin of the valvula foraminis ovalis and

the muscular part is not fused to any extent with the fibrous septum. The atrial septum as a whole is not in the same plane as the ventricular septum. Its anterior attachment is in line with that of the ventricular septum, but its posterior line of attachment is displaced toward the right, so that the mouth of the coronary sinus which is located just to its right, lies, not above the medial cusp of the tricuspid, as in the normal heart, but above the posterior cusp. The posterior line of attachment of the atrial septum is 2 cm. to the right of that of the ventricular septum.

In the right atrium neither the valve of the coronary sinus nor the valve of the inferior vena cava is to be found. An extremely delicate fibrous cord, attached to the atrial wall just below and lateral to the mouth of the sinus, and above to the superior wall between the two venae cavae, is seen hanging free in the atrium. This may possibly represent the remains of an undeveloped right sinus valve.

Interpretation

From our knowledge of the development of the heart, there can be no doubt that the malformation described depends upon two departures from the normal developmental processes: failure of fusion of the endocardial cushions, and persistence of the interventricular foramen. However, embryologists are not entirely agreed as to certain details in the normal course of these processes, so that before attempting to explain the anomalous features of this heart on the basis of these disturbances, the subject must be discussed in some detail. What seems fairly certain is that these disturbances date back to the first two months of intra-uterine life, for according to Mall [3] the development of the septa, ending with the closure of the interventricular foramen, is complete at an embryonal length of about 16 to 18 mm.; the embryo according to Arey [4] attains a crown-rump length of 17 mm. by the end of the seventh week.

On the subject of fusion of the endocardial cushions, and obliteration of the foramen ovale I, Mall differs from Tandler [5] and from Mönckeberg in one fundamental respect. According to the last two authors the interatrial septum (primum) grows downward to meet the endocardial cushions and fuses with them; Mönckeberg particularly emphasizes the presence of the lower margin of the atrial septum and upper margin of the ventricular septum, at the level of

the atrial canal, as a prerequisite for the fusion of the endocardial cushions. They form, according to this author, a sort of bridge across which the cushions may travel toward each other. In cases of patent foramen ovale I (primary interatrial foramen) with normal valves, the ventricular septum alone performs this function. He attributes the persistence of the primitive undivided atrial canal to a primary growth failure of both septa. On the other hand, Mall, in describing the closure of foramen ovale I, assigns more extensive growth to the cushions and less to the atrial septum; indeed, according to him, the obliteration of this foramen is due to the continued growth and fusion of those parts of the cushions which project into it. In an embryo of 9 mm. he describes the fused cushions as a "cubical plug which blocks the center of the atrial canal"; he does not specifically state that the foramen ovale I in this embryo is patent, but at 11 mm. he states: "The complete union of the two cushions has obliterated the foramen ovale I." Whether fusion of cushions in the atrial canal antedates obliteration of the foramen, or is coincident with it, is not specifically stated, but certainly, according to Mall, the fusion of the cushions with the atrial septum, as well as obliteration of foramen ovale I, is due to growth on the part of the cushions, and the original atrial septum never extends down to the level of the atrial canal.

It seems to be generally accepted that the definitive atrial septum is formed of three elements, the septum primum, septum secundum and the lower portion of the left sinus valve (Tandler). After septum I is well developed and perforated to form foramen ovale II, septum II appears as a feeble elevation at the right of septum I, extending downward from the postero-superior wall of the atrium. Septum II, continuing to increase in height, extends forward along the superior wall of the atrium and then downward along its anterior wall, and finally backward to meet the forward projecting left valve of the sinus venosus. The union of these two forms the limbus. Mall, who does not describe the formation of the atrial septum, does not mention the septum secundum. To Mall, all the elevations participating in the division of the primitive atrium, atrial canal, and bulbus are but parts of the subendocardial tissue which he describes in considerable histologic detail; they are not sharply separated from each other. Thus he describes at 7, 8, and 9 mm., both before and at the time of the fusion of the endocardial

cushions, prolongations of the cushions which extend upward "to the sinus venosus and are blended with the connective tissue above it." Again at 11 mm. he describes the fused mass (endocardial cushions) as extending up to the "atrial septum on its dorsal side and to the left valve of the opening into the sinus venosus." It is impossible to avoid the conclusion that the prolongations into the right atrium from this mass of connective tissue represent at least the lower part of Tandler's septum secundum. The attempt to apply Tandler's terms to these prolongations is made because it seems to establish an early continuity between the endocardial cushions, especially the posterior, on the one hand, and the septum secundum and sinus valves, especially the left, on the other. This connection is of interest in the anomalous heart under consideration.

In this heart the muscular portion of the atrial septum is defective. The lower limb of the limbus is absent. We may conclude that that portion of septum II which should have grown downward and curved backward, and there fused with the left valve of the sinus venosus, failed to develop in the usual manner. Furthermore, since the derivatives of the right sinus valve are also absent, a failure of development of both sinus valves seems to be indicated. It is possible, of course, that the valves of the coronary sinus and inferior vena cava were obliterated by the great dilatation of the right atrium, but more probably they were poorly developed, and hence left no trace in the atrium unless the fibrous cord mentioned above represents the right valve. If, as we have attempted to show, septum II of Tandler is either formed entirely, or increases in size, by the addition of those prolongations of Mall into the right atrium, then, failure of development of septum II and at least retarded development of the sinus valves, particularly the left, would in turn be dependent upon underdevelopment of the cushions, particularly the posterior. That in this heart the posterior cushion was deficient, will be shown later.

Another factor, however, may have contributed to the failure of fusion of the left valve of the sinus venosus with the endocardial cushions; namely, the obliquity of the atrial septum. Since the mouth of the coronary sinus lies so far to the right of the ventricular septum, this distance may have prevented the valve from fusing with the prolongation from the cushions.

Septum I apparently attained its usual growth, for it forms the

fibrous interatrial septum of the heart, separating the atria throughout at least three-quarters of their extent. Moreover, the upper free margin of the valvula foraminis ovalis is seen from the left side in its usual location at the upper anterior portion of the atrial septum. If, as Mall believes, the closure of foramen ovale I is due to the growth of the anterior and posterior cushions, the persistent interatrial aperture must be due to deficient growth of these cushions. The only features pointing to defective development of septum I, are the numerous small foramina perforating the fibrous septum. These might well be due to the fact that septum I was not reinforced all around its margin in the usual manner by septum II.

The obliquity of the atrial septum and the abnormal position of the coronary sinus, seem to be due to too great displacement of the opening of the sinus venosus. Early in embryonic development the opening of the sinus into the common atrium is made smaller by a fold coming in from the left side, and separating sinus from atrium (Tandler). In this way the sinus comes to open into the right side of the atrium. Septum I then develops just to the left of the mouth of the sinus. In this heart, the mouth of the sinus seems to have been pushed farther to the right than usual. The displacement of septum I may in turn have been partially responsible for the inability of the cushions to extend into and close foramen ovale I.

Regarding the interventricular septum, embryologists generally agree that coincident with the fusion of the atrial septum and cushions, the muscular interventricular septum increases in height and the bulbar septum grows down to meet it. During and after the fusion of these two septa, the interventricular foramen is reduced in size by continuous down-growth of the cushions. On the details of the above process, however, as well as in terminology, the various authors differ materially; they especially disagree on the exact mode of final closure of the interventricular foramen. It would lead us too far to go into the details of the controversy in which are included the opinions of Tandler, Mönckeberg, Sato, Born and Mall. Essentially the opinions held by these authors represent views varying more or less widely from the original description of His. It appears, however, that the diminution in size of the interventricular foramen is attributed to upward growth of the ventricular septum or down-growths from the cushions; its final closure to the bulbar septum, the cushions, or both.

In the heart under consideration, the complete separation of the pulmonary artery and the aorta and the well developed upper anterior part of the ventricular septum speak for the normal development of the entire septum aortopulmonale. The numerous intertrabecular apertures discernible on probing may be regarded as evidence that the ventricular septum was somewhat underdeveloped. Mönckeberg regards these as remnants of the spongy trabeculae composing the septum. The large aperture is, however, not much larger than a normal membranous septum would be, and it seems quite probable that it was somewhat increased in size during dilatation of the heart. We conclude, therefore, that the ventricular septum was not greatly deficient in its growth.

Certain structures found in the place of the membranous septum and apparently derived from the anterior and posterior cushions, seem to indicate the part played by the cushions in the formation of the septum. For example, the chordae and fibrous leaf which attach the finger-like mass of endocardial tissue to the posterior margin of the aperture, may represent that process from the posterior cushion which, according to Mall, normally grows downward along the posterior margin of the foramen. Inasmuch as the chordae attaching the anterior cusp of the mitral valve to the anterior margin of the septum occupy the same position as the membranous septum of a normal heart, it would seem that they represent cushion material which normally goes into the formation of the membranous septum. In a recent report of a heart similar to this, Schleussing [6] describes a small fibrous area lying "in the normal location of the pars membranacea" (between right and posterior semilunar aortic cusps?) and believes that this part of the septum membranaceum is formed by the bulbar septum. In our heart a similar small fibrous sheet is found, but it attaches the aortic cusp to the base of the right semilunar cusp, and is clearly similar to the chordae tendineae lying immediately below it. Our heart seems, therefore, to offer some evidence in favor of the theory advanced by Sato [7] that the cushions are entirely responsible for the closure of the interventricular foramen.

Regarding the development of the atrioventricular valves, the original description of His seems, on the whole, to stand unchallenged. As to the exact part played by the endocardial cushions, both medial and lateral, in valve formation, His, Sato, and Mall

seem to be the only authors who have fully discussed this subject. Mall, who agrees with His, shows by the study of a graded series of human embryos that the medial cusp of the tricuspid is derived from the right ends of both cushions, and that the anterior medial papillary muscle is formed by the fusion of the septum aortopulmonale with the medial end of the right lateral cushion. It, therefore, marks the boundary between the anterior and right lateral cushions, and may be used in the interpretation of anomalous valve formation. Sato's theory of valve formation has usually been the accepted one, namely, that the right end of the anterior cushion takes part in the formation of the anterior cusp of the tricuspid.

It should be stated, in favor of the Mall theory, that Mall actually finds at one stage of development, the medial cusp of the tricuspid and the aortic cusp of the mitral hanging like curtains over the septum; this points toward the derivation of the medial cusp, like the aortic, from both endocardial cushions, and not as Sato believes, from the right end of the posterior only. Mall finds, also, in numerous embryos of less than 20 mm., that the two venous ostia are quite alike; each is bordered by a medial and lateral cusp, and each ventricle has a papillary muscle or set of muscles between the medial and lateral cusps. Finally, Mönckeberg describes in a trilocular heart, with single atrium but completely formed ventricular septum and atrioventricular cusps, the two medial cusps (aortic of the mitral, and medial of the tricuspid) continuous with each other over the top of the septum.

In this heart, the aortic cusp of the mitral is obviously derived from the left end of the anterior cushion, and is, therefore, the equivalent of that half of the normal aortic cusp which lies farthest from the septum. The small strand of tissue connecting the aortic cusp with the anterior of the tricuspid is, in the light of Mall's theory, derived from the right end of the anterior cushion; like the anterior end of the normal medial cusp, it is attached to the anterior medial papillary muscle of the right ventricle. The right end of the anterior cushion may have been more deficient in its growth potentialities than the left end. It may, however, have remained small secondarily, for all the cusps lacked the stimulus for development which would have been supplied had the cushions fused. Mall describes an attachment between the septum aortopulmonale and the right end of the anterior endocardial cushion similar to that be-

tween the septum and lateral cushion. The only attachment acquired by the right end of the anterior cushion (to the septum aortopulmonale) tended, therefore, to draw it to the left, rather than out into the right ventricle and further hindered its increase in size.

As to the posterior cushion, the small medial cusp of the tricuspid was formed from its right end, and is, therefore, from Mall's standpoint, the equivalent of only the posterior half of the normal medial cusp. The posterior cushion failed to send a prolongation down into the left ventricle; consequently there is no cusp to correspond to the septal half of the aortic; in turn, no posterior papillary muscle was developed in the left ventricle. The posterior cushion material which should have gone into the aortic cusp is represented by the finger-like process. Marked underdevelopment of the posterior cushion is indicated by the character of its derivatives.

The right lateral cushion and the heart wall posterior to this cushion gave rise to the anterior and lateral (posterior) cusps of the tricuspid. The left lateral cushion gave rise to the small lateral cusp found in the left ventricle, which is the only representative in this heart of the normal posterior cusp of the mitral. The remainder of the posterior cusp, which should have been derived from the heart wall posterior to the lateral cushion by a process of undermining, was probably hindered in its development by the absence of the posterior papillary muscle.

The above interpretation of the anomalous valves, based on Mall's description of the embryologic development of the heart, seems to us most acceptable. Another interpretation, based on the theory of Sato, is, however, possible. According to his theory, the anterior cusp of the tricuspid is normally formed from the right end of the anterior cushion plus the major part of the right lateral cushion. In the light of this, the small strand of valve tissue which connects the aortic cusp of the mitral with the anterior of the tricuspid, represents, not the right end of the anterior cushion, but only that part of the cushion which lay above the interventricular septum. If this theory is correct, the anterior cushion was quite normal and the failure of fusion of the two cushions would have to be attributed almost entirely to growth failure of the posterior cushion.

Summary

The essential features of the heart described are as follows:

1. A large aperture of communication between the two sides of the heart, involving the lower atrial and the upper ventricular septa.

2. A fibrous interatrial septum, the posterior attachment of which, together with the coronary sinus, is displaced to the right.

3. A common atrioventricular canal bordered by five cusps — in the left ventricle, a small lateral and a large anterior, and in the right, a large anterior and small lateral and medial. The lateral corresponds to the posterior of the normal tricuspid valve.

4. A mass of endocardial tissue attached to the posterior margin of the interventricular septum, and projecting into the interventricular aperture.

It is obvious that the defects in this heart are the result of failure of fusion and imperfect development of the endocardial cushions, incomplete formation of septum II, and displacement of the mouth of the sinus venosus to the right. If Mall's description of the mode of closure of the foramen ovale I is accepted, the persistence of this foramen must be due to primary growth deficiency on the part of the cushions, rather than of septum I, as is commonly held. If the views of Mall regarding the relations of the lower portions of the left sinus valve are reconciled with those of Tandler, as we have attempted to do, it appears that imperfect formation of septum II may also be due to growth deficiency of the cushions. We have shown how the abnormal valves of this heart could be interpreted either in terms of Mall's or of Sato's descriptions of valve formation. Finally, since much of the site of the normal membranous interventricular septum is occupied by chordae tendineae, this heart seems to offer some evidence that the cushions normally play an important part in, if indeed they are not entirely responsible for, the closure of the interventricular foramen.

Case 2

Clinical Report: The patient was a female child, aged four months, who died in the Children's Hospital (No. 2776). Physical examination showed Mongolian facies, malnutrition, cyanosis, a loud murmur over the precordium, and a short fifth digit. The Wassermann reaction was positive. The necropsy took place three and one-half hours after death.

Anatomic Diagnoses: (1) Defect of the atrial septum, with patent foramen ovale; defect of the upper part of the ventricular septum; common atrioventricular canal with abnormal atrioventricular valves. Patent ductus arteriosus.

(2) General hypertrophy and moderate dilatation of the heart especially of the right auricle. (Measurements in the fixed condition: apex to base, 4.3 cm.; transverse diameter, 3.8 cm.; circumference at base, 11.0 cm. Weight: 33 gm.)

(3) Mongolian facies.

(4) Purulent bronchitis and lobular pneumonia.

Description of the Heart

The muscular interventricular septum, which in its thickest portion measures 0.9 cm., has a concave upper margin which forms the lower boundary of the interventricular aperture. The aperture measures 1.5 cm. in an anteroposterior direction, and 0.7 cm. from the line of attachment of the atrioventricular valves to the free margin of the septum.

The upper anterior portion of the septum differs somewhat from that of the preceding heart. A muscular process from the anterior part of the septum extends backward and upward under the right aortic cusp to the posterior cusp. In the normal heart, as well as in the preceding, the area just under the point of junction of the right and posterior cusps is fibrous, forming in the normal heart, the uppermost part of the membranous septum; in this heart, this region is muscular.

Probing between the trabeculae of the ventricular septum reveals a number of points of communication between the ventricles.

The valve cusps surrounding the common atrioventricular canal are five in number — two anterior, which are continuous with each other through the aperture; a single posterior, common to both ventricles; and one lateral in each ventricle.

The papillary muscles of the left ventricle are normal, namely, an anterior and a posterior. In the right ventricle the posterior medial muscle may be considered normal. The anterior medial is represented, as in many normal hearts, by a group of chordae attached to the crista supraventricularis, or posterior wall of the conus; of this group, only one is fleshy. The anterior (large anterior) papil-

lary muscle is rather small. The moderator band, extending from the vase of the anterior papillary muscle to the ventricular septum, is unusually large in this heart, being thicker than the anterior papillary muscle itself, and short enough to hold the anterior and septal walls of the right ventricle in close apposition.

In the left ventricle there is a large cusp corresponding to the lateral half or two-thirds of the aortic cusp of the mitral. It arises from the lower margin of the adjacent halves of the left and posterior aortic cusps. Its lateral margin is attached to the anterior papillary muscle of the left ventricle. The upper part of its medial margin is attached by chordae tendineae to the anterior portion of the upper curved margin of the ventricular septum and to that muscular process which extends backward under the right aortic cusp. This attachment is made by chordae, which are not, however, completely separated from each other but are united into a sheet by a delicate web of endocardium stretching between them. From the lower part of the medial margin of the aortic cusp, chordae pass across the top of the septum to attach to the right surface of the septum.

The cusp just described is continuous by means of a narrow strand of valve tissue with the anterior cusp in the right ventricle. This strand is attached by chordae to the free margin of the septum in its upper anterior portion, and to the crista by means of some of the chordae (including the only fleshy member of the group) which we have called collectively, the anterior medial papillary muscle. The remainder of the chordae comprising this group attach the medial border of the anterior cusp of the right side to the crista supraventricularis; the lateral border of the same cusp is attached to the anterior papillary muscle. This cusp arises from the annulus fibrosus lateral to the conus. The lateral cusp in the right ventricle, which corresponds to the posterior of the normal heart, is attached by chordae both to the anterior and to the posterior medial papillary muscles.

The posterior cusp is well developed, in fact rather redundant, and is common to both ventricles, extending across the septum. In the right ventricle it is attached to the posterior medial papillary muscle, to the right surface of the septum and to the posterior part of the upper margin of the septum. Of those chordae which are attached to the septum some extend well forward to obtain attach-

ment to a rather prominent muscular projection from the moderator band. In the left ventricle the posterior cusp is attached to the left posterior papillary muscle. The lateral cusp in the left ventricle is narrow, and is attached to both anterior and posterior papillary muscles.

The atrial septum of this heart is somewhat better developed than that of the preceding. Though the disposition of the muscular portion is similar, ending below on the anterior wall of the right atrium, it comprises rather more of the atrial septum than in the preceding case. The fibrous portion (valvula foraminis ovalis) is thicker and not perforated. As in the preceding heart, its lower free margin forms the upper boundary of the interatrial aperture. The upper curved margin of the fibrous portion of the septum, falls just short of being overlapped by the muscular portion, thus forming a patent foramen ovale II. This septum is nearly in the same plane as the ventricular septum, its posterior attachment being displaced only very slightly to the left, instead of markedly to the right, as in the preceding case. Neither the valve of the coronary sinus nor of the inferior vena cava is present. As in Case I, there is a great discrepancy in size between the two atria and auricles (auricular appendices), the right being very much larger than the left. This difference is especially marked in the auricles.

Interpretation

Since the heart in this case is in all essential features similar to the one previously described, the interpretation given for that heart will apply to this one. However, there are a few points of difference which must be discussed. The shifting of the mouth of the sinus venosus to the right, which in the preceding case, was assumed to explain the obliquity of the atrial septum, apparently does not occur in this heart, for the mouth of the coronary sinus lies directly above the right side of the ventricular septum. The derivatives of the posterior endocardial cushions are not so conspicuously underdeveloped as in the preceding. Both cushions are deficient, however, as compared to the normal.

The well developed muscle connecting the septal and anterior walls of the heart, which we have called the moderator band, presents difficulties. Mönckeberg believes that the septum aorticum

(*i. e.*, probably that portion called swelling A by Tandler), as it grows down to meet the ventricular septum, divides into two limbs, one passing to the left, and the other to the right of the ventricular septum. The limb which passes to the right, carries the right branch of the atrioventricular bundle downward, and forms the moderator band. As was noted, the posterior cusp in this heart, obviously derived from the posterior cushion, is attached by chordae to a small papillary projection on the moderator band. For an explanation of this, three possibilities suggest themselves: (1) That Mönckeberg's theory of formation of the moderator band is incorrect; (2) that the muscle in question, in spite of its location, is not actually the moderator band; (3) that the posterior cushion, growing down into the right ventricle, obtained an accidental attachment to the moderator band. The latter possibility is not inconsistent with Mall's description of the formation of papillary muscles and chordae tendineae; nor do the chordae in question extend farther forward on the septum than chordae attaching the posterior half of the medial cusp of the tricuspid to the septum in many normal hearts.

Brief Review of Similar Cases from Literature

Preisz [8] describes two cases, both in new-born children. In neither was the membranous atrial septum (valvula foraminis ovalis) as well developed as in these hearts; in both, the atrial canal was surrounded by four valve cusps, a large anterior and posterior, and two small laterals. In one case the anterior cusp was attached to the anterior limb of the interventricular septum; in the other, this attachment was absent.

Hart [9] reports two cases, one aged two years, and the other ten months. Both showed large anterior and posterior atrioventricular valve cusps, whose chordae attached in the right ventricle. One of the cases, however, showed complete absence of the atrial septum. It is, therefore, a trilocular heart with ventricular septum defect, and does not strictly belong in the same class as the hearts under discussion. It has not been included in the total.

Mönckeberg describes one case in a child of nine months. The atrial septum was fibrous and perforated to form a foramen ovale II; its lower margin reached to a distance of 1 cm. from the atrioventricular canal; the septum secundum was entirely absent. The

common atrial canal was guarded by large anterior and posterior cusps (lateral cusps are not mentioned); and under the posterior aortic cusp was located a mass of endocardial tissue about the size of a grape seed. The midportion of the crest of the ventricular septum, forming the lower margin of the aperture, was free from valve tissue, as in our cases.

Schleussing[6] reports two cases, one in a female Mongolian idiot of nine and one-half months, and another in a new-born child. The first, in which the atrial septum was similar to the first of our hearts, was complicated by some degree of transposition of the great vessels, and by the fact that the medial limb of the bulbar septum passed to the right of the ventricular septum. The valve cusps were five in number, a large anterior, continuous across the ventricular septum, two laterals and two posteriors. In the second case the atrial septum showed a large foramen ovale overlaid anteriorly and above on the right side by a perforated membranous fold. The cusps were four in number, a large anterior and posterior, and two laterals. He found in both cases a structure which he interpreted as the pars membranacea.

Keith[1] states that he has seen fourteen cases, all remarkably alike. He does not consider that the atrial septum was defective, though his figure shows a condition like that of the hearts described here. Unless the atria show a much larger aperture than in these cases he does not regard it as a persistent foramen ovale I. He states that failure of fusion of the endocardial cushions is always associated with some other grave defect, such as transposition of the great vessels or stenosis of the pulmonary artery; but in only two cases other than Keith's have defects of the great vessels been reported. In one of Preisz's cases the pulmonary valve had only two cusps; in one of Schleussing's cases there was partial transposition and narrowing of the aorta.

Keith also cites two cases similar to his, described by Griffith.

II. Common Atrium, Completely Divided Ventricles, and Imperfectly Formed Tricuspid Valve

This type of heart, with completely separated ventricles and common atrium, is called by Mönckeberg, *cor triloculare biventriculosum.*

Case 3

Clinical Report: The heart is that of a male child, who died at the age of two months, in the Children's Hospital (No. 1971). The child was one of twins, born prematurely at the end of the seventh month of pregnancy. The only findings at necropsy, other than the abnormal heart, are general atrophy of the organs, hemorrhages in the pleura, lungs and capsules of the kidneys, and a marked slant of the palpebral fissures characteristic of Mongolism.

Description of the Heart

The heart is approximately normal in size and presents no abnormalities externally. When it is opened, the two atria are observed to be in wide communication. The atrial septum is represented only by a low muscular fold which extends from the left side of the coronary sinus to the lower margin of the defect just mentioned. The muscular fold extends forward for about two millimeters along the top of the membranous septum, forming the posterior part of the lower boundary of the aperture. The anterior part of the boundary is formed by the line of fusion of the membranous septum with the aortic cusp of the mitral valve.

The ventricular septum is completely formed. Its membranous portion is considerably larger than usual. The latter lies below the interatrial defect, and has a triangular process extending backward below the rudimentary atrial septum. On the right side, the membranous septum is not covered by the tricuspid valve as in the normal heart, because of malformation of this valve; a few small nodular thickenings appear on this surface. The top of the membranous septum is continuous with the medial third of the aortic cusp of the mitral, the two merging into each other without interruption.

Both cusps of the mitral and the papillary muscles of the left ventricle may be considered normal. The cusps of the tricuspid present certain malformations. Both they and the papillary muscles in the right ventricle are smaller than normal. With the exception of the posterior, the cusps are nodular and adherent to the septum or wall of the heart against which they lie. The posterior cusp is

quite free; it arises from the posterior and lateral portions of the atrioventricular junction, and sends numerous fine chordae tendineae to the posterior medial papillary muscle and to the adjacent portion of the septum. The medial cusp is deficient in its anterior one-third; it arises from the upper margin of the muscular portion of the interventricular septum. It is adherent to the septum and might be described as a patch of nodular and thickened endocardium on that portion of the septum. Its lower margin, however, is free, and from this margin chordae pass to the posterior medial papillary muscle and to the septum. It has also a single chorda passing to the lowest point of junction of the membranous septum with the muscular septum.

Between the membranous septum and the crista supraventricularis there is a crumpled mass of valve tissue from which two groups of chordae pass, one to the surface of the membranous septum itself and to the lowest point of junction of membranous with muscular septum, and the other to the lowest point of the crista. The latter group of chordae are gathered together and are attached to a single, fairly well developed papillary muscle which constitutes the major portion of the anterior medial muscle.

The crumpled mass just described is directly continuous with the anterior cusp of the tricuspid. This cusp is fairly smooth, but, like the medial, is closely adherent to the heart wall, excepting that portion which joins it to the crumpled mass. From this junctional piece, a single chorda passes across the crista and is attached within the conus to its posterior wall. This chorda and the single papillary muscle mentioned above, comprise the anterior medial papillary muscle. The chordae of the anterior cusp pass to the large anterior papillary muscle. The latter is poorly developed, consists of two trabeculae, and lies against the lateral (anterior, in the normal position of the heart) wall of the ventricle.

Interpretation

That septum I was deficient or entirely absent in this heart seems clear. Because of its location medial to the mouth of the coronary sinus, the rudimentary atrial septum, which is muscular, seems to have been derived from the left valve of the sinus venosus. It corresponds to the lower and posterior part of the normal limbus of the fossa ovalis. Its continuity with the posterior part of the mem-

branous septum recalls the early fusion between an upward prolongation of the posterior cushion in the right atrium, and the left valve of the sinus venosus (Mall).

The cusps in the right ventricle will be interpreted first in the light of Mall's theory of valve formation. As noted previously, the anterior medial papillary muscle marks the boundary between the anterior and right lateral cushions. Examination of a series of normal hearts, has shown that both medial and anterior cusps are usually attached to the muscle and that it may consist of several parts, either muscular or tendinous. In the heart under discussion, it consists of a group of chordae attached to a fairly well developed papillary muscle, and a single chorda passing into the conus. The point between these two is taken as approximately the line of demarcation between the anterior and lateral cushions. The crumpled mass, therefore, represents the right end of the anterior cushion. Since this mass is distinctly separated from the medial cusp of the tricuspid, the latter, in this heart, seems to have been derived from the right end of the posterior cushion alone. The right ends of the cushions apparently failed to fuse.

Another interpretation, based upon the theory of valve formation as given by Sato, is possible. Sato holds that the normal medial cusp of the tricuspid is derived from the right end of the posterior cushion alone, and that the right end of the anterior cushion goes into the anterior cusp. The crumpled mass, if regarded as part of the medial cusp, must be a part of the posterior cushion, split off secondarily; if regarded as belonging to the anterior cusp, it is the right end of the anterior cushion. In the latter case, it must be assumed that the growth potentialities of the posterior cushion were insufficient to form a normal medial cusp.

In a recent study of malformations of the heart, Mönckeberg has no cases exactly similar to this one. He describes, however, one case of *cor triloculare biventriculosum*, in which there was no vestige of the atrial septum, and in which the apex was cleft.

Summary

The heart just described presents, in addition to almost complete absence of the atrial septum, defects of the tricuspid valve, especially its medial cusp. The defect of the atrial septum is due to absence

or degeneration of the septum primum, and to absence of the septum secundum. The rudimentary atrial septum is interpreted as the left valve of the sinus venosus. The defective medial cusp of the tricuspid is interpreted as due either to failure of fusion of the right ends of the endocardial cushions (if Mall's description is accepted), or to deficient growth potentialities of the posterior cushions alone (according to the theory of Sato).

REFERENCES

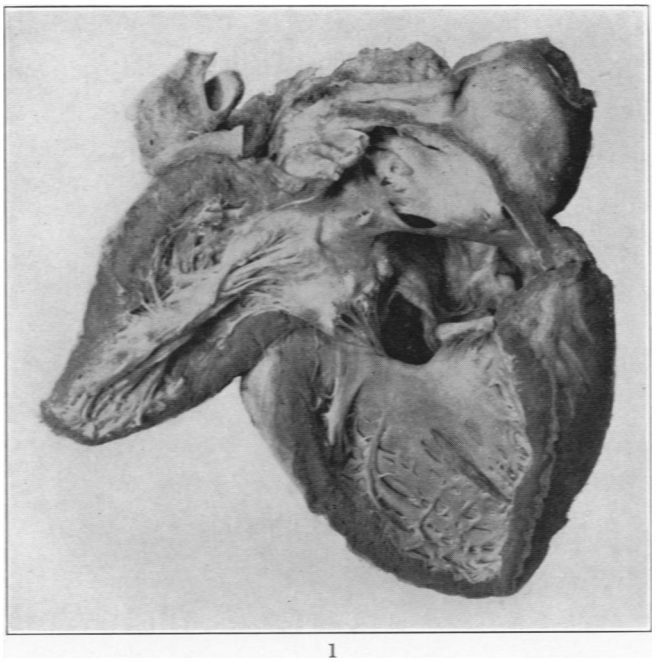
1. Keith, A. The Hunterian lectures on malformations of the heart. *Lancet*, 1909, ii, 359, 433, 519.
2. Mönckeberg, J. G. Die Missbildungen des Herzens, Handbuch der speziellen pathologischen Anatomie und Histologie. Henke und Lubarsch, 1924, ii.
3. Mall, F. P. On the development of the human heart. *Am. J. Anat.*, 1912, xiii, 249.
4. Arey, L. B. Developmental Anatomy. Philadelphia, 1924, 68.
5. Tandler, J. Development of the Heart. Keibel, F. K., and Mall, F. P., Human Embryology. New York, 1912, ii.
6. Schleussing, H. Beiträge zu den Missbildungen des Herzens. *Virchows Arch. f. path. Anat.*, 1925, ccliv, 579.
7. Sato, cited by Mönckeberg.
8. Preisz, H. Beiträge zur Lehre von den angeborenen Herzanomalien. *Zieglers Beitr. z. path. Anat.*, 1890, vii, 245.
9. Hart, C. Ueber die Defekte im oberen Teilder Kammerscheidewand des Herzens, mit Berücksichtigung der Perforation des häutigen Septums. *Virchows Arch. f. path. Anat.*, 1905, clxxxi, 51.

DESCRIPTION OF PLATES

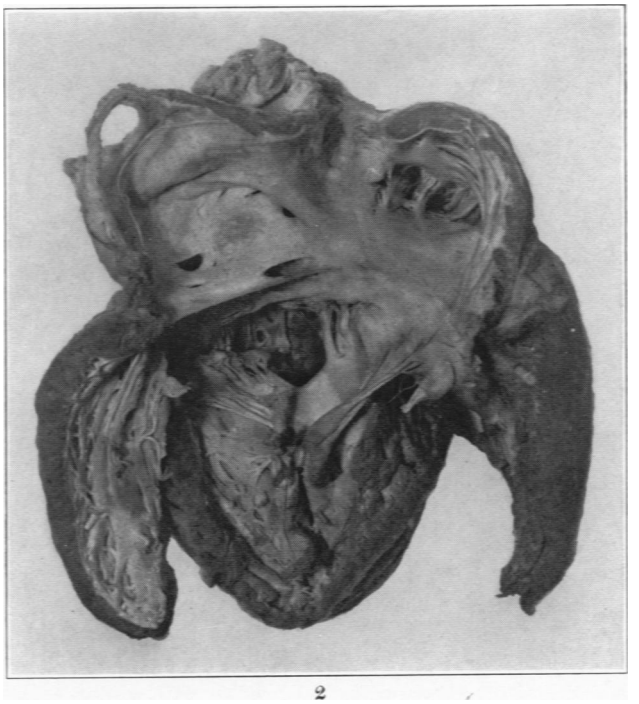
PLATE 156

FIG. 1. Left atrium and ventricle of heart from Case 1. The finger-like process projecting into the aperture from the posterior heart wall is seen to the right of the aperture.

FIG. 2. Right atrium and ventricle of heart from Case 1. The finger-like process is here seen projecting into the aperture from the left.



1



2

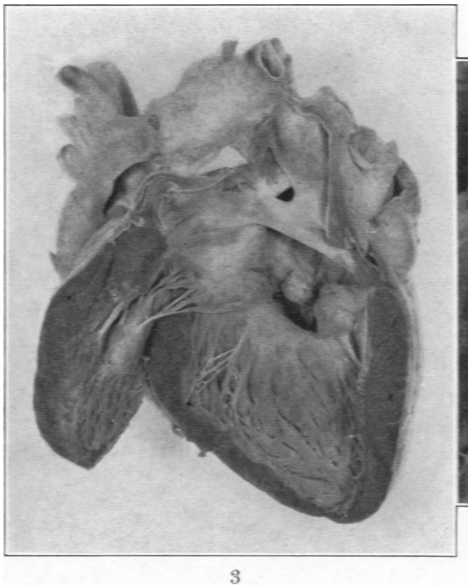
PLATE 157

FIG. 3. Region just below the aorta from the heart of Case 1. The attachment to the base of the right semilunar cusp is by a small sheet of fibrous tissue, and to the anterior margin of the aperture by chordae tendineae.

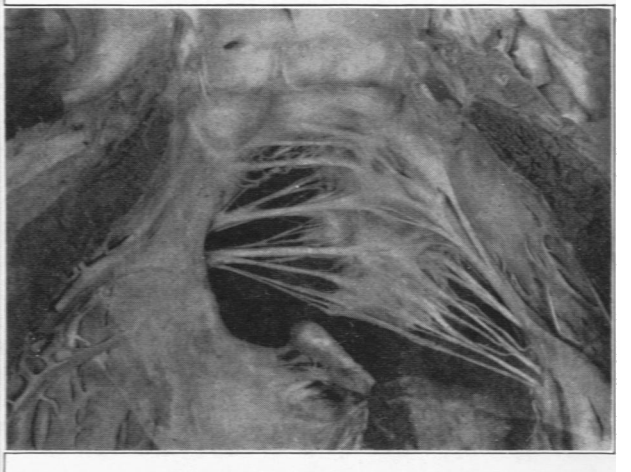
FIG. 4. Left atrium and ventricle of heart from Case 2.

FIG. 5. Right atrium and ventricle of heart from Case 2.

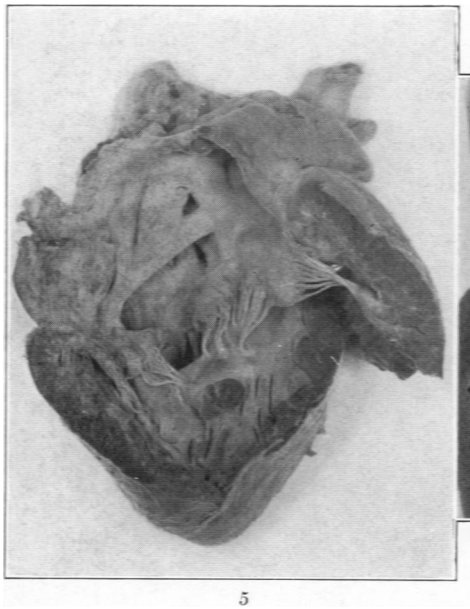
FIG. 6. Region just below the aorta from the heart of Case 2. The attachment to the muscular process that extends backward under the right semilunar cusp as well as to the upper part of the anterior margin of the aperture is by chordae tendineae, which are joined together by a delicate web of endocardial tissue. The lower chordae pass through the aperture to the right side of the top of the septum.



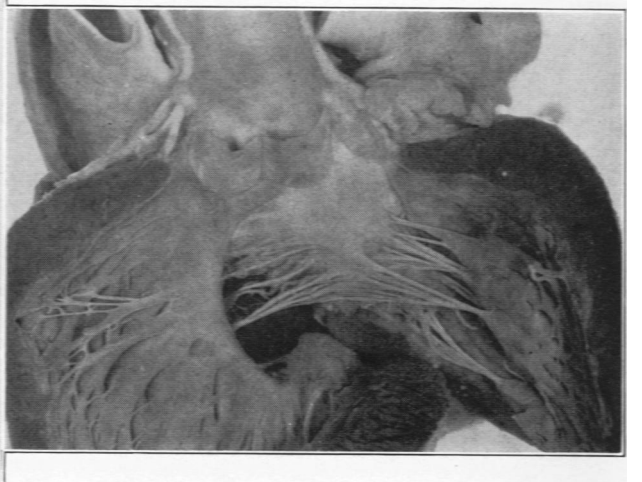
3



4



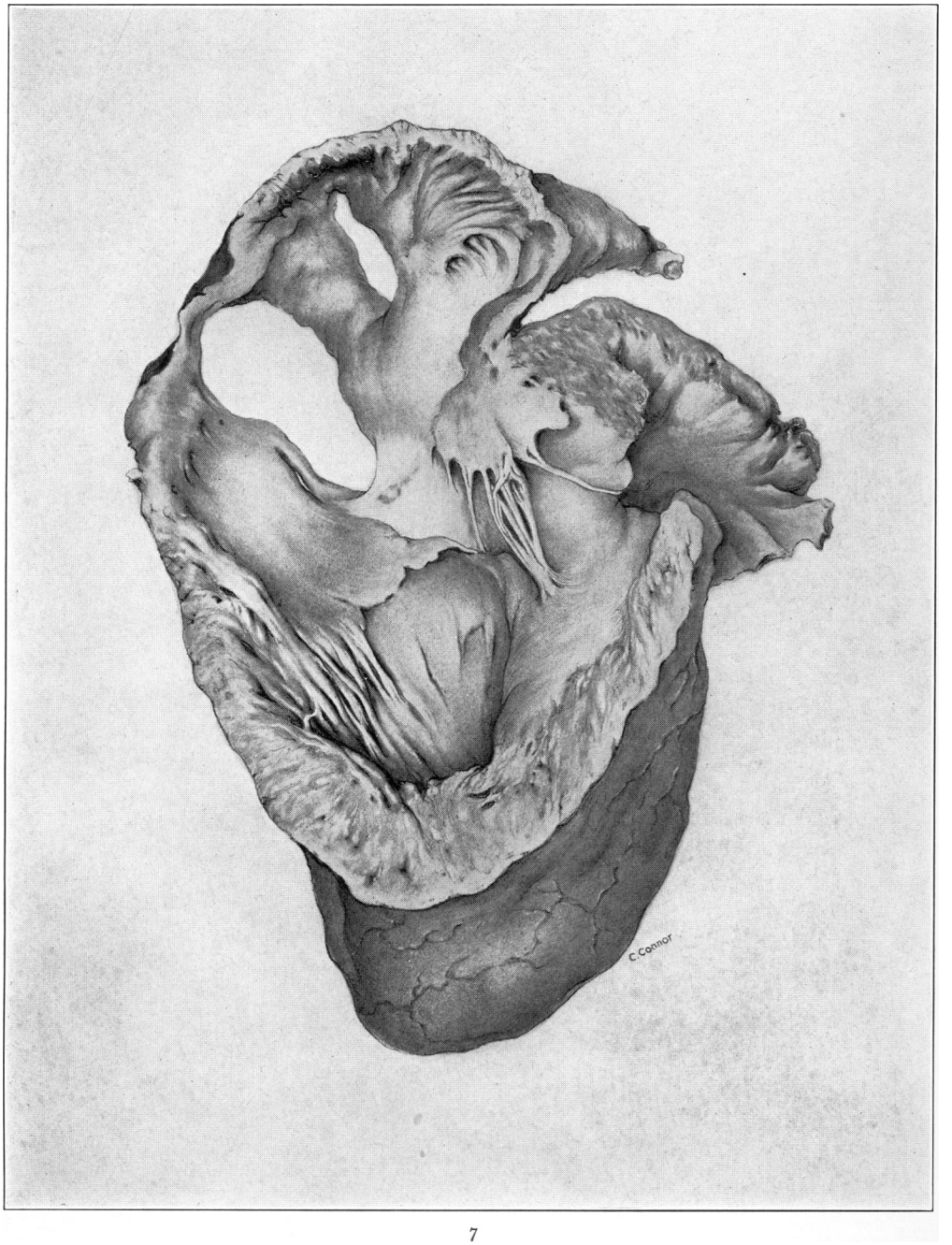
5



6

PLATE 158

FIG. 7. Right atrium and ventricle of heart from Case 3, the anterior wall of the heart removed.



7