REPORT OF AN UNUSUAL CASE OF CONGENITAL CARDIAC DISEASE, DEFECT OF THE UPPER PART OF THE INTERAURICULAR SEPTUM (PERSISTENT OSTIUM SECUNDUM), WITH, FOR COMPARISON, A REPORT OF A CASE OF PERSISTENT OSTIUM PRIMUM.

By MAUDE E. ABBOTT, M.D., and
JOSEPH KAUFMANN, M.D.

Reprinted from The Journal of Pathology and Bacteriology.
Vol. XIV. (1910).

Journal of Pathology and Bacteriology, Vol. XIV.

REPORT OF AN UNUSUAL CASE OF CONGENITAL CARDIAC DISEASE. DEFECT OF THE UPPER PART OF THE INTERAURICULAR SEPTUM (PERSISTENT OSTIUM SECUNDUM),

WITH, FOR COMPARISON,

A REPORT OF A CASE OF PERSISTENT OSTIUM PRIMUM.[1]

By MAUDE E. ABBOTT, M.D., *Governor's Fellow in Pathology;* and JOSEPH KAUFMANN, M.D., *Assistant-Curator of the Pathological Museum, McGill University, Montreal.*

(PLATES XXXI.–XXXIV.)

CASE 1 (Plates XXXI.–XXXIII. Figs. 1, 2, and 3).—The unique specimen which forms the subject proper of this paper was presented to the Pathological Museum of McGill University by Dr. F. W. C. Mohr of Ottawa, by whose kind permission the case is reported The following brief clinical notes have been received through the courtesy of Dr. Cousins, under whose observation the patient was during life.

A. B., a woman, æt. 64, married, had worked very hard as a charwoman nearly all her life. Until six years before death she had perfect health, but since that time she had noticed that at times she was not as well as usual, and that occasionally her lips and finger-tips became blue. She was admitted to hospital on 10th July 1908, in a dull semi-stuporous state, suffering from extreme dyspnœa, the lips and finger-tips deeply cyanosed, the face only slightly less so, pulse imperceptible at the wrist, the lower extremities extremely œdematous, the abdomen distended with fluid, the heart dulness enlarged both to left and right. Death occurred a few hours after admission, before a physical examination of the chest had been made. No data of interest other than the cardiac anomalies were noted at the autopsy.

DESCRIPTION OF SPECIMEN.

SUMMARY.—*Large defect of the upper and posterior part of the interauricular septum above the foramen ovale, apparently due to defective development of the secondary auricular septum. Absence of the Eustachian valve and of annulus ovalis. Dilatation of the tricuspid orifice and thickening and incompetency of the tricuspid valves. Chronic sclerosing endocarditis*

[1] Received January 21, 1910.

with enlargement, thickening, distortion, insufficiency, and calcification of the pulmonary valves. Widening of the pulmonary orifice and marked dilatation of the pulmonary artery. Great hypertrophy and dilatation of the right auricle and ventricle. Hypoplasia of the aorta and of aortic vestibule of left ventricle.

The specimen is an adult heart of unusually large size and of peculiar contour, the enlargement being confined almost entirely to the right chambers, so that the apex is formed by the right ventricle, and the right auricle rises high above the base; while the large right auricular appendix projects well forward to the right of the pulmonary artery, the small left appendix being barely visible anteriorly. The epicardial fat is increased. The great vessels have been cut away near their base, and the condition of the ductus arteriosus is therefore not known; the great veins have also been cut away close to the auricles, so that an abnormality in the course of the pulmonary veins, a common associated anomaly in auricular defects in this situation, can unfortunately neither be determined nor excluded.

The interauricular septum presents a large ovoid defect obliquely placed in its upper and posterior part, $3 \times 3{\cdot}5$ cms. in diameter, which is bounded postero-superiorly by the roof of the auricles, which are here smoothly continuous with each other, while its antero-superior, anterior and lower borders are formed by the defective interauricular septum, the free margin of which is thick and rounded in front, but below is thin and sharply curved. The septum itself is a thick muscular partition 1·5 to 2 cms. high, which rises upward and backward from the interventricular septum, with which it is continuous below; it is a massive muscular structure throughout, except along its upper border (the lower border of the defect), where it becomes very thin, the muscular portion appearing as though surmounted by a falciform fold of endocardium placed more towards the left than the right, which seems to represent the remains of the fossa ovalis, and which presents on the side of the left auricle a slit-like blind pocket, apparently the closed foramen ovale. On the side of the right auricle the muscular structure of the septum sends forward a heavy limb dividing the coronary sinus from the inferior vena cava. *There is, however, no trace of the annulus ovalis*, of which this ridge would be the termination in the normal heart, nor of the Eustachian valve, which should surmount it. The defective septum is placed further to the left than to the right, cutting off a large right and a small left auricle.

The right auricle is much dilated, and is also greatly hypertrophied with heavily developed musculi pectinati, and an appendix of huge size. It receives two veins; the smaller of these, evidently the superior vena cava, opens into the roof of the auricle close to the defect, but is separated from this by a thick muscular ridge which extends upwards to the vault of the auricle from the anterior portion of the defective septum; the lumen of the superior cava appears to look, not towards the defect, but downward and forward into the cavity of the auricle, as in the normal heart; the larger venous opening, that of the inferior vena cava, lies in the posterior wall of the right auricle a little below and 4 cms. to the right of the superior cava. The coronary sinus, which is much dilated, lies 3 cms. below the orifice of the inferior cava; it is separated from the latter by the thick muscular ridge mentioned above, which runs to it from the defective auricular septum, which ridge in the normal heart is continuous with the anterior cornu of the annulus ovalis on the one hand and the Eustachian valve on the other; *but here both of these structures are wanting.* The coronary sinus is guarded by an imperfect valve abnormally placed, and, close to its mouth, receives two dilated veins from the anterior surface of the right ventricle. *No pulmonary veins enter the right auricle.*

The right ventricle is dilated and greatly hypertrophied (Plate XXXI. Fig. 2), presenting a remarkable compensatory development of its musculature, so that its trabeculæ with the greatly enlarged moderator band and the papillary muscles of the tricuspid together form a heavy bridge-like structure, which seems to divide off the conus from the sinus of the ventricle, making the former appear almost like a third chamber communicating with the sinus by a rounded orifice formed by the arching muscle-bundles. The trabeculæ are slightly flattened, as from pathological dilatation. The conus is of enormous size, and is somewhat altered in shape with a very short posterior wall (1·5 cm.). The tricuspid orifice is dilated, 13 cms. in circumference, the valves are thickened, and the infundibular cusp heavily developed.

The pulmonary orifice is wide, the pulmonary cusps are much *enlarged, thickened, and stiffened, and the two anterior are the seat of extensive coral-like calcareous excrescences*, the result of a deposit of lime salts in the fibrosed valve tissue (Plate XXXII. Fig. 2). The pulmonary artery is much dilated, 9 cms. in circumference, and is thick-walled, but presents no signs of atheroma.

The left auricle is small, about a quarter the size of the right. One pulmonary vein enters the roof posteriorly close to the defective septum ; but, owing to the fact that the auricle had been laid open rather irregularly before the specimen was seen, no other opening into its walls could be detected, and as all the vessels had been cut off close to their entrances the course of the other pulmonary veins could not be traced (Fig. 3).

The left ventricle is smaller than the right. The endocardium is fibrosed, and the columnæ carneæ are small and atrophic, contrasting markedly with those of the right ventricle. The papillary muscles are plump, not flattened. The mitral orifice is 8 cms. in circumference and the valves appear normal. The aortic valves are normal and competent, but the orifice is small and the aortic vestibule of the ventricle is narrow. *The aorta itself is very small, measuring only 5 cms. in circumference*; it presents some patches of atheroma about the sinuses of Valsalva (Plate XXXIII. Fig. 3).

Defects of the interauricular septum, other than a simple patency of the foramen ovale, are not common, and when they do occur they are usually at the *lower* part just above the interventricular septum, so that both auricles communicate with each other and with the auriculo-ventricular ostia by an ovoid opening placed *below* the fossa ovalis (*persistent ostium primum*).

This form of defect is that shown in the following case, which is here reported and figured (Plate XXXIV. Fig. 4) for the sake of comparison.

CASE 2.—*Defect of the lower part of the interauricular septum of the heart (persistent ostium primum). Malformation of tricuspid and mitral valves, with cleavage of the anterior mitral segment. Hypertrophy and dilatation of both auricles, especially of the right. Hypertrophy and dilatation of the conus arteriosus of the right ventricle and dilatation of the pulmonary artery. Presented to the Pathological Museum of McGill University by Professor J. G. Adami.*

From a strong well-developed man, æt. 35, who died of perforative appendicitis the day after admission to the Royal Victoria Hospital. Some irregularity of the heart's action was noted, and roughening of the first sound at the apex, but no cyanosis.

The specimen is an adult heart with a broad base. The foramen ovale is completely closed, but the lower part of the interauricular septum is wanting,

a crescentic aperture through which two fingers can be passed with ease, existing. This opening is bounded above by the thin concave edge of the valvula foraminis ovalis; below, it impinges directly on the interventricular septum, its lower border being formed on the side of the left ventricle by projecting cushions sent upward from the base of the two halves into which the anterior mitral segment is divided. The right auricle is much enlarged, the fossa ovalis is increased in size and presents irregular thickening, but is nowhere perforated; the annulus ovalis is absent below opposite to the defect, and does not curve forward to meet the Eustachian valve. The sinus of the right ventricle is of about normal size, but its conus arteriosus is much enlarged and has very thick walls; it is quite cut off from the sinus by the thickened and very strong infundibular cusp of the tricuspid valve, which stretches obliquely across it. The anterior chordæ of this cusp arise from the greatly hypertrophied anterior papillary muscle, while its posterior ones run horizontally back to a heavy band of muscle, which passes from the posterior (septal) wall of the ventricle to the lower margin of the defect in the interauricular septum, a gap 15 mm. long existing at this point between the septal and infundibular tricuspid cusps.

The anterior segment of the mitral valve is thickened and sclerotic, and is divided into two halves by a cleavage along its middle; each half runs upward in an oblique direction from its papillary muscle to the lower margin of the defect, where the anterior half overlaps the posterior, a triangular interval existing between them below and giving the appearance at first sight of an additional cusp. *This deformity of the anterior mitral segment is present in the majority of cases of defect of the lower part of the interauricular septum (persistent ostium primum), and was seen in five of Rokitansky's six cases.*

The left auricle is dilated and hypertrophied; the left ventricle is of about normal size.

The pulmonary artery is about one-third larger than the aorta.

Discussion.

Defects at the *upper* part of the septum, *above* the fossa ovalis, as in the specimen which forms the subject of this paper, are exceedingly rare. Rokitansky, it is true, describes nine cases of persistent *ostium secundum* in his great work on defects of the Cardiac Septa (1875[1]), but only six others are to be found in the literature after an extended search. These are by Wagstaffe (1868 [2]) (two cases), Chiari (1880 [3]), Hepburn (1887 [4]), Greenfield (1890 [5]), Ingalls (1907 [6]), and Ellis (1906 [7]). Our specimen is unique among those recorded in (*a*) the age of the patient, which is the highest attained, and (*b*) the association of a primary chronic pulmonary endocarditis from overwork with extreme calcareous change.

The following analysis of the recorded cases of defect at the upper part of the interauricular septum is of interest in considering both the pathogenesis and the clinical significance of the abnormalities in the present case.

Wagstaffe.

Case 1.—A child, æt. 6. In good health until the onset of scarlet fever ten weeks before death. A circular opening, 6 lines in diameter, seen best on the side of the right auricle, lay in the upper part of the septum atriorum

above the foramen ovale, which was valvular. Right carotid and subclavian rose from the arch.

CASE 2.—Female, æt. 52, dying of acute pericarditis, heart sounds indistinct from pericardial effusion. Large opening in the upper part of the auricular septum directly below entrance of superior vena cava, 1 in. across, lining membrane of auricles smoothly continuous with each other over the edge of this. Foramen ovale closed. Aortic and mitral stenosis. Dilatation of pulmonary and tricuspid orifices. Great enlargement of right chambers, especially of auricle. Ductus arteriosus closed.

Rokitansky.

CASE 7.—Male, æt. 19. Œdema of extremities and cyanosis in last 14 days of life. Pleurisy with effusion. Ovoid defect in upper and back part of auricular septum 32 by 39 mm. in diameter, bounded below and anteriorly by the rudimentary auricular septum, 21 mm. high, which is thick and muscular in front, but behind is thin and falciform, and contains here a patent foramen ovale. *Orifice of inferior cava looks into both auricles, right pulmonary veins open into right auricle, pulmonary artery much dilated, hypoplasia of the aorta.* Enlargement of heart, especially of right chambers and conus. Passive congestion of organs.

CASE 8.—Female, æt. 50. Auricular septum absent except for a ridge 13 mm. high above the septum ventriculorum, from which a fleshy bundle passes upward to the roof of the auricles dividing the orifice of the superior cava from the pulmonary veins. *Dilatation of pulmonary artery.* Enlargement of heart, especially of right chambers. Thrombosis of pulmonary artery. Pericarditis.

CASE 9.—Female, æt. 20. In place of the upper part of the auricular septum is a large hole bounded by a muscular ridge which is prominent below, becomes smaller above and anteriorly, and is almost imperceptible behind. *Pulmonary dilatation.* Enlargement of right heart. Adherent and calcified pericardium.

CASE 10.—Male, æt. 50. Œdema of extremities and some cyanosis of body. Large defect 40 mm. wide bounded by a fleshy frame which is prominent below (21 mm. high) anteriorly and above, but is almost obliterated behind, and has a membranous lower free border. *Pulmonary dilatation. Ostium of vena cava inferior looking into both auricles,* marked enlargement of right heart, thickening of auriculo-ventricular cusps.

CASE 11.—Female, æt. 23, dying from pulmonary tuberculosis. Defect of auricular septum at upper and back part 33 mm. in diameter, bounded below and anteriorly by a fleshy ridge which has a membranous free border. *Pulmonary artery dilated. Orifice of inferior cava looks into both auricles.* Enlargement of right heart.

CASE 12.—Male, æt. 44. Defect of auricular septum 35 mm. wide, bounded by a fleshy framework which is 12 mm. high below, above, and in front, and is very low posteriorly, and has a membranous free border that *sends a limb to surround the orifice of the inferior vena cava, so that this looks directly into the left auricle. Pulmonary dilatation.* Enlargement of right heart.

CASE 13.—Male, æt. 35. Accidental death. Defect of auricular septum 33 mm. in diameter, bounded by a muscular ridge that is very prominent below, becomes shallower above and anteriorly, and is obliterated behind. The lower part of this ridge becomes very thin and membranous, and runs backward to divide the orifice of the inferior vena cava from the coronary sinus (Eustachian valve), *causing the former to look directly into the left auricle through the defect.* A delicate membranous cord runs across the defect, arising at the border of the ostium of the inferior vena cava, and is inserted anteriorly

into the muscular substance of the defective septum on the side of the left auricle. *Aorta narrow, pulmonary artery very wide.* Enlargement of right heart.

CASE 14.—New born child. Defect of auricular septum 15 by 17 mm., bounded below and behind by a fleshy partition and containing a very delicate floating network which is inserted on the side of the left auricle but passes to the outer border of the orifice of the inferior vena cava and becomes continuous with a similar network in front of the ostium venæ coronariæ (Thebesian valve). *Congenital pulmonary stenosis*, enlargement of right heart, patent ductus arteriosus.

CASE 15.—Male, æt. 21. Defect of auricular septum 29 mm. in diameter, bounded on its lower and anterior aspects by a fleshy ridge 8 mm. high, and traversed on the side of the left auricle by delicate threads and by a membrane perforated in the form of a cross which projects into the left auricle, *into which the orifice of the inferior cava partly looks.* Dilatation of pulmonary artery. *Hypoplasia of aorta.* Enlargement of right heart.

CASE 16.—Male, æt. 43. Round defect of auricular septum 35 mm. wide, bounded by a fleshy ridge which is 11 mm. high below, 17 mm. high above and anteriorly, and is obliterated behind. Ostium of inferior vena cava looks partly into left auricle. A fine network arises from the upper part of the defective septum and extends into the orifice of the superior vena cava on the one hand, and on the other passes backward into the left auricle, forming here an obliquely placed diaphragm, extensively perforated, which is stretched above the left auriculo-ventricular orifice as an *anomalous septum. Pulmonary dilatation.* Enormous enlargement of the heart, especially of the right auricle.

Chiari.

Opening in upper part of septum atriorum 15 mm. in diameter. Foramen ovale patent. *One large and three small pulmonary veins open into right auricle to right of a line connecting superior and inferior cavæ.* Enlargement of heart, especially of right ventricle.

Hepburn.

Adult male, subject from dissecting-room. *Upper right pulmonary vein enters superior vena cava close to right auricle*, and two lower right pulmonary veins are firmly adherent to the posterior wall of the right auricle, but empty into the left close to the septum. Ovoid foramen in upper part of septum atriorum, one inch above and quite distinct from the upper margin of the fossa ovalis; its short axis measures three-eighths of an inch, its long axis is in the line of the superior vena cava, its upper end being close to the posterior margin of the opening into the right auricle common to the superior vena cava and the upper right pulmonary vein. Foramen ovale closed. *Persistent left superior cava.*

Greenfield.

Railway porter, æt. 53. Had done much heavy lifting, and had been a heavy drinker all his life. Always healthy until eighteen months before death, when failing compensation with œdema and cyanosis set in. Improved under rest, etc., but he had two subsequent attacks; in the second of these, which was characterised by marked cyanosis, he died suddenly. Examination of the heart on the first admission showed enlargement, especially to the right, slight presystolic thrill at the apex, purring presystolic murmur at fourth left costal cartilage (heard later also at the third), transmitted downward to the apex, where also there is a short blowing systolic murmur transmitted to the axilla, and the second sound is loud and clear, sometimes reduplicated; rough

blowing systolic murmur at aortic, loud systolic at pulmonary cartilage, aortic and pulmonary accentuation. The autopsy revealed a circular defect in the auricular septum 1½ in. in diameter, bounded below and posteriorly by a septum, and above by the wall of the auricles. *Patent foramen ovale* about half inch below the defect. Right pulmonary veins were cut away, and their point of entrance was not known. *Superior vena cava placed over the defect, so that blood passed into both auricles. Bicuspid aortic valve, stenosis of aortic orifice. Pulmonary dilatation.* Enormous hypertrophy and dilatation of right chambers, moderate of left heart. *Coarctation of the aorta.* Pleurisy with effusion.

Ingalls.

Adult male. No cardiac symptoms. Heart slightly enlarged, chiefly on the right side. *Two upper right pulmonary veins enter the superior vena cava.* Fossa ovalis well defined, foramen ovale not patent. Valvula Eustachia well developed. High up in the auricles the wall of the septum is deficient, so that the left auricle communicates with the superior vena cava just where this enters the right atrium. The defect is limited below by the smooth, deeply concave margin of the defective septum, the free edge of which blends posteriorly with the wall of the vena cava superior *just below the entrance* of the medial right pulmonary vein, while its other extremity can be traced on the antero-medial wall of the superior vena cava as far as a point opposite the centre of the vena pulmonalis superior dextra, thus dividing the cava into two parts, of which that looking into the left auricle is the larger. The defect is not well seen from the right auricle.

Ellis.

Female, æt. 32. An opening in the anterior part of the auricular septum 32 mm. in diameter. A thin place in the membrane of the fossa ovalis below and behind this represents the closed foramen ovale.

From a perusal of the above cases it will be seen that certain anomalies are commonly associated with defects in the upper part of the interauricular septum, and one or other of these has been argued by different authorities to be primary to the septal defect. Thus in several instances (Chiari, Hepburn, Rokitansky's Case 7, Ingalls, and probably Greenfield's) the right pulmonary veins were displaced, and opened either into the superior vena cava or into the right auricle. The presence of the additional blood from this source in the right auricle was thought by Chiari to be the cause of the septal defect. In the cases of Ingalls and Hepburn, in which not only were the right pulmonary veins displaced, but the superior cava rode over the defect in the septum looking into both auricles, the defect was considered to be due to a development too far to the right of the septum primum, so that it came to be directly under the opening of the superior vena cava. This explanation cannot apply in the present specimen, in which the superior vena cava clearly enters the right auricle, and is separated from the defect and from the pulmonary vein entering the left auricle by a steep muscular cushion derived from the defective septum (Plate XXXI. Fig. 1); here also no pulmonary veins enter the right auricle, though whether any of these vessels enter the superior vena cava it is impossible to say, as this has

been cut off short, and the roof of the left auricle mutilated so that the position and number of the veins entering this chamber cannot be determined.

The explanation given by Rokitansky and cited below seems to apply best in the present case. Indeed, it seems possible that this specimen, with his Cases 8 to 16, and with that of Greenfield, falls pathogenetically into one category; while his Case 7 and those of Ingalls, Hepburn, and Chiari, in all of which there is a displacement of the right pulmonary veins, lie in another. The auricular septum is known to develop in two parallel planes (Born, His), which grow downward from the roof of the common auricle at a very early stage. Of these two, that developing first, the *septum primum*, lies on the left side; as it grows downward an opening remains for some time at its lower border above the interventricular septum. This is known as the *ostium primum*, and the persistence of this orifice is seen in Case 2 (Plate XXXIV. Fig. 4). Gradually, as the primary septum grows downward, this *ostium primum* closes in, and a second opening appears above, in the upper part of the primary septum. This is the *ostium secundum*; it assumes a valvular appearance by the growth downward of the *septum secundum*, which begins on the roof of the auricle to the right of the primary septum and arches downward to form the *annulus ovalis* of post-natal life. Rokitansky considers a defect in the upper part of the auricular septum to be, in most cases, a persistence of the *ostium secundum* due to a non-development of the secondary septum. In other cases he thinks it may be an anomalous opening above the foramen ovale due to non-closure of some of the fenestrations present in the primary septum at a still earlier stage of development. He also draws attention to the fact that nearly all the cases of this defect are associated with a hypoplasia of the aorta and dilatation of the pulmonary artery, and suggests that this is congenital and the narrowing of the aorta is the primary condition, the increased tension in the left auricle setting in directly after birth and causing widening of a small congenital slit at the upper margin of the auricular septum.

In our case the absence of an annulus ovalis or Eustachian valve argues for a true developmental absence of the secondary septum; the hypoplasia of the aorta is a prominent feature; while the presence of the primary chronic pulmonary endocarditis with calcification, together with the extreme hypertrophy of the right ventricle, indicates an increased amount of blood in the right chambers to be expelled into the pulmonary circulation, leading in its turn to raised peripheral pressure there, and to consequent overwork of the pulmonary valves. The sources of this excess of fluid in the right chambers are, blood entering from the left auricle through the defect, blood regurgitating through the insufficient pulmonary valves, and possibly also blood entering the right auricle from displaced pulmonary veins

opening into the superior vena cava. The peculiar development of the trabeculæ in the sinus of the right ventricle suggests an attempt to resist an influx of blood through the defect.

Clinical Aspects.

Defects in the upper part of the septum seem to have little clinical significance, and usually present no evidence of their presence during life. In Greenfield's case alone was a presystolic murmur heard over the fourth left interspace, a sign usually considered characteristic of an auricular septal defect, observed. The fact that the patients were practically all adults, and that in our case, as also in that of Greenfield, a long life of unusual strain was undergone without any evidences of ill-health or cyanosis until shortly before death, indicates that under ordinary circumstances there can have been but little interference with the conditions of the normal circulation; that is, there was either little or no admixture of currents through the defect, or else (and this is supported by Rokitansky's view) a current passed only from left to right through the defect, arterial thus mixing with venous blood. In the following four cases only did a terminal cyanosis develop: In our patient, in whom the cyanosis was intermittent during the last six years of life, finally becoming intense; in that of Greenfield, in whom it was noticed in the last eighteen months during three successive attacks of failing compensation; in Rokitansky's Case 7, cyanotic only in the last fourteen days, and in Rokitansky's Case 10, in whom "some cyanosis" is mentioned. In none of the other cases was cyanosis observed. These facts argue that with the development of an embarrassed pulmonary circulation from the increased amount of fluid in the right heart and its consequent overwork, the pressure in the right auricle was increased and the current through the defect reversed, venous blood being carried through it to the left auricle and thence to the systemic circulation. These features place this case (and the three above mentioned) with the *cyanose tardive* of the French authors (1889 [8]).

From the evidence of many recorded cases it is definitely seen that the system can accommodate itself to a certain amount of unaerated blood entering directly into the arterial stream from some abnormal source. As soon, however, as this admixture has passed a certain limit, which happens when pulmonary embarrassment supervenes and the admixture of venous blood is increased by the rise of pressure in the right ventricle and the consequent passage of an increased quantity of blood from the right into the left auricle through the defect (as seems to have been the case here), malaeration becomes extreme, and the symptom-complex of congenital cyanosis develops.

Conclusions.

From an analysis of the literature and the study of Case 1, the following facts are elicited:—

1. Defects at the upper part of the interauricular septum are among the rarest of cardiac anomalies. Anatomically there appear to be two groups of cases: those associated with displacement of the pulmonary veins, and those in which no such anomaly is present. In nearly all the cases recorded, hypoplasia of the aorta was a prominent feature.

2. Defects in this situation, even when very large, are usually latent throughout life, giving rise to no physical signs, and being unassociated with cyanosis; in the cases in which cyanosis was present this was terminal.

3. This case especially shows that the existence of a large defect in the upper part of the interauricular septum is compatible with a long life of even more than the average stress; nevertheless, the changes in the right heart and the advanced disease of the pulmonary valve were significant of increased strain in the pulmonary circulation, which probably led to the final breakdown.

REFERENCES.

1. Rokitansky . . . . . "Die Defeckte der Scheidewände des Herzens," Vienna, 1875.
2. Wagstaffe . . . . . *Trans. Path. Soc. London*, 1868, vol. xix. p. 96.
3. Chiari[1]. . . . . *Jahrb. f. Kinderh.*, Leipzig, 1880, Bd. xv. p. 319 (quoted by Ingalls).
4. Hepburn . . . . . *Journ. Anat. and Physiol.*, London, 1887, vol. xxi. p. 438.
5. Greenfield . . . . . *Ibid.*, London, 1890, vol. xxiv. p. 423.
6. Ingalls. . . . . *Johns Hopkins Hosp. Bull.*, Baltimore, April 1907, p. 136.
7. Ellis . . . . . *Proc. Path. Soc. Philadelphia*, N.S., 1906, vol. ix. p. 44.
8. Bard et Curtillet. . . *Révue de Méd.*, Paris, 1889, tome ix. p. 993.

[1] Not verified.

DESCRIPTION OF PLATES XXXI.–XXXIV.

Plate XXXI.

Fig. 1.—*Heart from Case* 1, *showing defect in the upper part of interauricular septum* (*persistent ostium secundum*), *with pulmonary dilatation, sclerosing pulmonary endocarditis with calcification, and hypoplasia of the aorta.* (*From a woman æt.* 64.)

Right chambers laid open to show:—

(*a*) Hypertrophied and dilated right auricle.

(*b*) Large defect in upper part of interauricular septum, bounded below by

(*c*) A stout muscular partition, the defective auricular septum.

(*d*) Absence of annulus ovalis and of Eustachian valve.
(*e*) Dilated coronary sinus guarded by a defective valve of Thebesius.
(*f*) The greatly hypertrophied and dilated sinus of the right ventricle.
(*g*) Entrance of superior vena cava.
(*h*) Entrance of inferior vena cava.
(*i*) Passage to conus of right ventricle.

(From a specimen in the Pathological Museum of M'Gill University, Montreal.)

PLATE XXXII.

FIG. 2.—*Heart from Case* 1. *View of the conus arteriosus of the right ventricle, showing its great hypertrophy and dilatation, the great dilatation of the pulmonary artery and orifice, and the marked thickening, deformity, and calcification of the pulmonary valves.*

(*a*) Superior vena cava entering the right auricle.
(*b*) The dilated pulmonary artery.
(*c*) Calcareous excrescences on pulmonary valves.
(*d*) The hypertrophied and dilated conus of the right ventricle.
(*e*) The communication of the conus with the sinus of the right ventricle.

PLATE XXXIII.

FIG. 3.—*Heart from Case* 1. *The left chambers laid open to show* :—

(*a*) The relatively small size of the left auricle as compared with the right.
(*b*) The defective interauricular septum surmounted posteriorly by a thin fold of endocardium, containing :—
(*c*) The closed foramen ovale.
(*d*) A pulmonary vein entering the left auricle close to the defect.
(*e*) The back of the dilated right auricle.
(*f*) The small aorta, and narrow aortic vestibule.
(*g*) The cut edge of the dilated pulmonary artery.
(*h*) The superior vena cava entering the right auricle.
(*i*) The defect in the upper part of the auricular septum.
(*j*) The mitral valve.

PLATE XXXIV.

FIG. 4.—*Heart from Case* 2, *showing large defect in the lower part of the interauricular heart* (*persistent ostium primum*). *Dilatation of the pulmonary artery, and cleavage of the anterior segment of the mitral valve.* (*From a man æt.* 35.)

Left chambers laid open to show :—

(*a*) The dilated left auricle.
(*b*) The fossa ovalis with closed foramen ovale.
(*c*) Defect of the lower part of the interauricular septum (persistent ostium primum).
(*d*) Cleavage of anterior segment of mitral valve.

(From a specimen in the Pathological Museum of M'Gill University, Montreal.)

b. Defect in auricular septum.

h. Inferior vena cava.

d. Muscular ridge Eustachian valve and Annulus ovalis absent.

e. Dilated coronary sinus.

f. Sinus of right ventricle.

g. Superior vena cava.

c. Interauricular septum.

i. Passage to conus.

Fig. 1.

R.A.
Aorta
P.A.
R.V.
a. Superior vena cava.
b. Dilated pulmonary artery.
c. Calcareous excrescences on pulmonary valve.
e. Communication with sinus of ventricle.
d. Hypertrophied conus of R.V.

Fig. 3.

a. Superior vena cava.

d. Pulmonary vein.

c. Defect in A.S.

c. Closed foramen ovale.

b. Defective auricular septum.

f. Mitral valve.

g. Pulmonary artery.

f. Sarron aortic vestibule.

Fig. 2.

a. The dilated left auricle.

b. Fossa ovalis.

c. Defect of inter-auricular septum below.

d. Anterior segment of mitral valve, showing cleavage.

Fig. 1.

d. Anterior segment of mitral valve, showing cleavage.