EBSTEIN'S MALFORMATION OF THE TRICUSPID VALVE A REPORT OF SIX CASES

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Ebstein's malformation consists of a deformed tricuspid valve arising in whole or in part from the wall of the right ventricle below the annulus fibrosus at the atrioventricular junction, with the result that the portion of the ventricle proximal to the abnormal valvular attachment forms with the right atrium a large common receiving chamber. The right ventricle is therefore reduced in size and consists mainly of the anatomic outflow portion of this chamber. An associated atrial septal defect or patent foramen ovale is usually present and may give rise to a venous-arterial shunt with resultant cyanosis.

The first case was described by Ebstein in 1866, but it was not until 1949 that a diagnosis was made in a living patient with the aid of cardiac catheterization (Tourniaire et al.). Since then, due to a better understanding of the clinical picture and the more widespread use of modern diagnostic methods, many reports of single cases or series of cases recognized during life have been published (Reynolds, 1950; Soloff et al., 1951; Blacket et al., 1952; van Lingen et al., 1952; Henderson et al., 1953; Gotzsche and Falholt, 1954, Medd et al., 1954; Kerwin, 1955; Brown et al., 1956; Blount et al., 1957; Mayer et al., 1957; Hunter and Lillehei, 1958; Kezdi and Wennemark, 1958; Schiebler et al., 1959; Sinha et al., 1960). Two recent publications (Kilby et al., 1956; Vacca et al., 1958) review the literature up to date, recording a total of 122 reported cases.

Although no specific treatment is available, the diagnosis of this congenital cardiac defect is of more than academic interest. Failure to recognize the condition has in the past led to a mistaken diagnosis and surgical intervention with fatal results (Engle et al., 1950; Baker et al., 1950; Gasul et al., 1953; Lev et al., 1955), as occurred with our first patient. Furthermore, cardiac catheterization in patients suffering from this condition is not without danger and has often resulted in fatal complications (Baker et al., 1950; Blacket et al., 1952; Campbell, 1953; Mayer et al., 1957); this indicates the need for correct diagnosis on clinical findings alone.

The purpose of this paper is to present six cases of Ebstein's malformation. Clinical observations were made in four of these patients, two of whom came to necropsy. Two others were not seen during life; they died suddenly at home and only post-mortem findings are available.

CLINICAL FEATURES

Case 1: A Chinese boy, aged 14 years, was first seen on November 28, 1953. He had been ill for four years with mild breathlessness on exertion and swelling of the abdomen and feet. The symptoms were not severe and he had been treated by various doctors as an outpatient. In April 1952 he was thought to be suffering from tuberculous pericardial effusion, was admitted to hospital for one month and given a course of streptomycin. After discharge from hospital, he was still not well but remained ambulatory. One week before his second admission his symptoms became severe.

His general condition became progressively worse in hospital and subsequently a thoracotomy was done under the mistaken impression that he was suffering from tricuspid stenosis. At operation, from which he did not recover, the pericardial sac was distended by about 600 ml. of clear yellow fluid and gross cardiac enlargement due to a large right atrium was found.

Case II: A 14-year-old Chinese girl was admitted on April 3, 1957 complaining of severe breathlessness, cough and swelling of the legs of seven days' duration. She had had similar complaints on and off for the past 1½ years, but the symptoms had been relatively mild. Although quite ill and orthopnoeic on admission, she improved considerably and felt fit enough to walk about the ward. Early one morning, a month after admission, she was found dead in bed.

Case III: A Chinese boy, 14 years of age, was referred to hospital on August 27, 1955 because on routine medical examination by the School Health Officer he had been noted to have a large heart. He had no complaints and his parents had not noticed anything abnormal about him. Although he said he became breathless when he played games, his exercise tolerance appeared quite good. When last seen about two years ago, he still had no symptoms and was attending school.

Case IV: A Chinese girl, aged 16 years, was seen on March 3, 1959 with the complaint

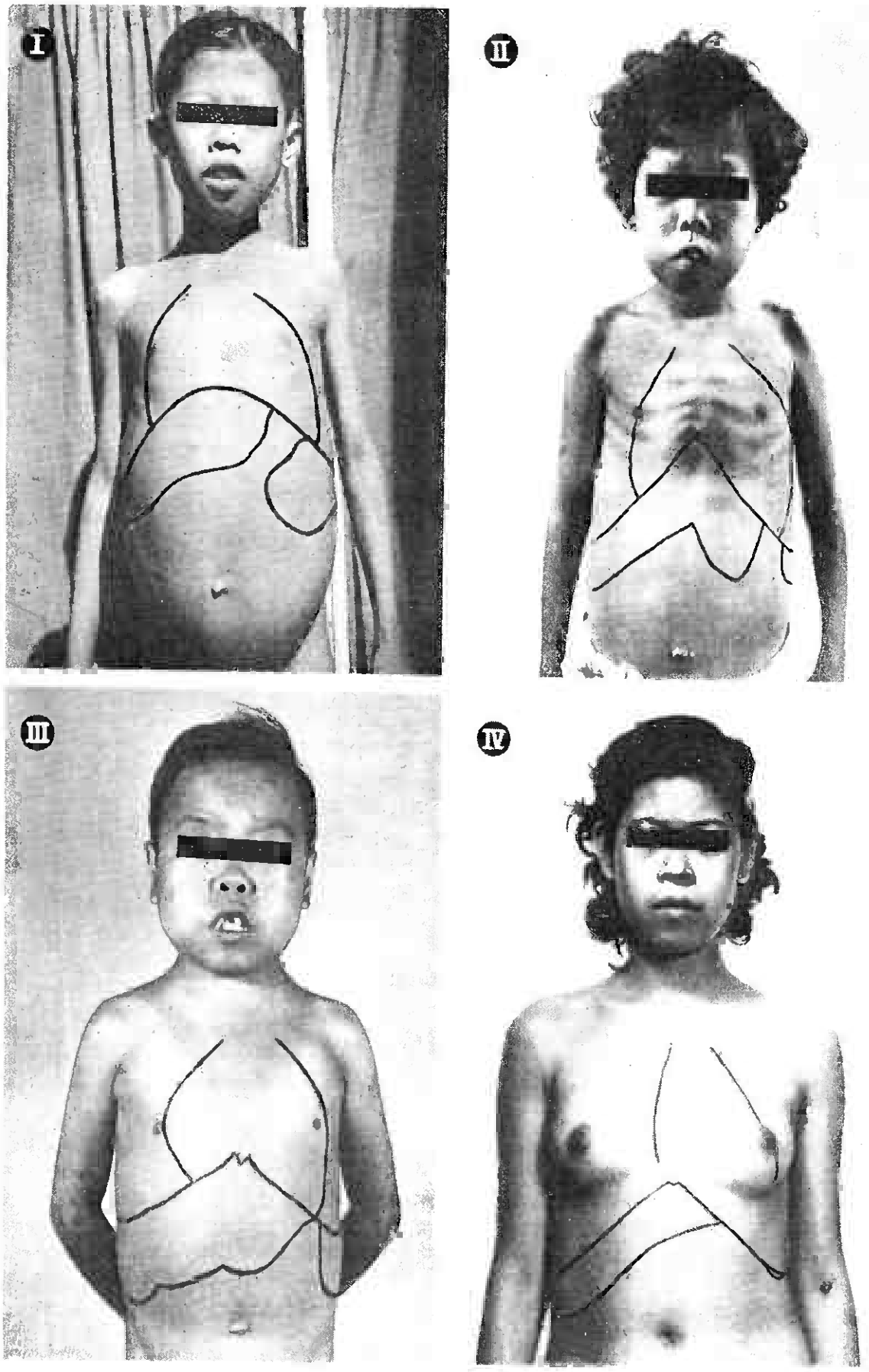

Fig. 1. Physical appearance of puffy face, cardiomegaly, hepato-splenomegaly and distended abdomen in Cases I-IV. Note the "moon facies" in Case III

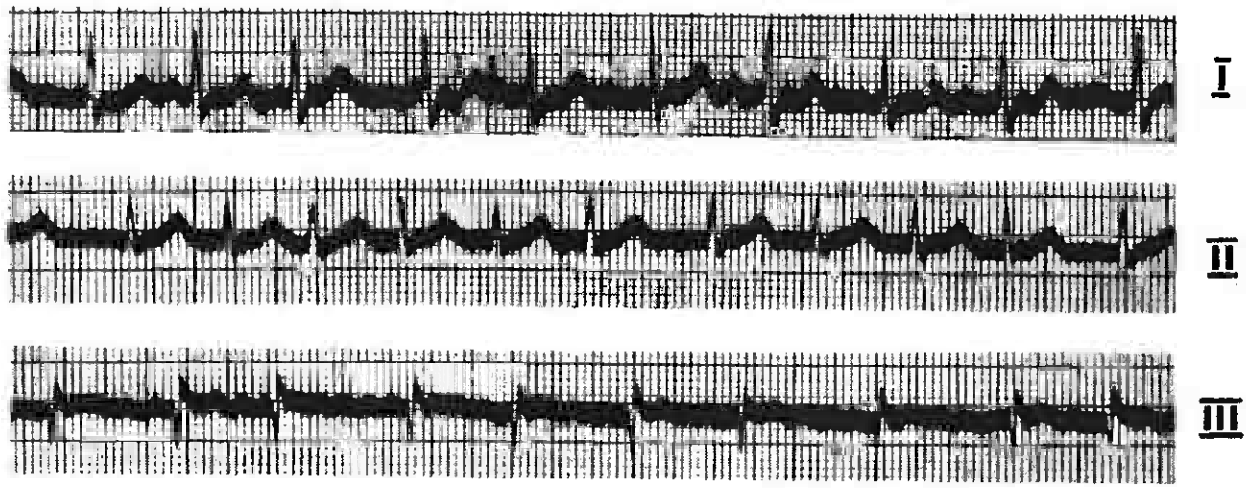

Fig. 2. Case III. Electrocardiogram showing auricular fibrillation.

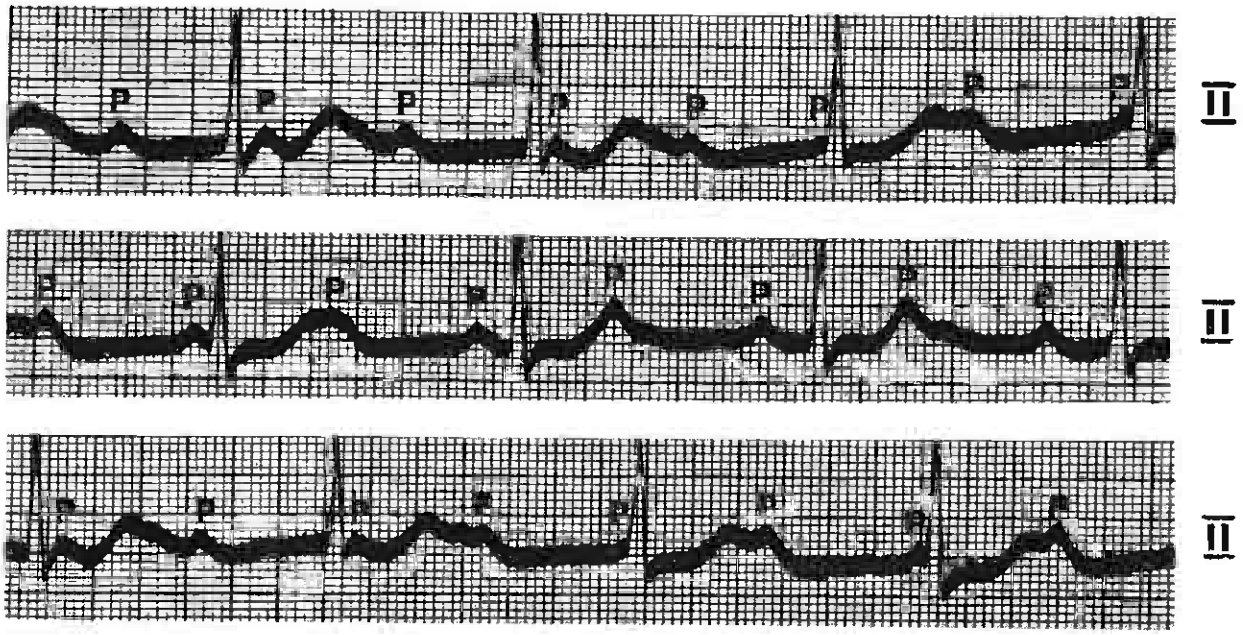

Fig. 3. Case IV. Electrocardiogram showing complete heart block. The standardization has been increased (1mv=30mm) so as to bring out the P waves.

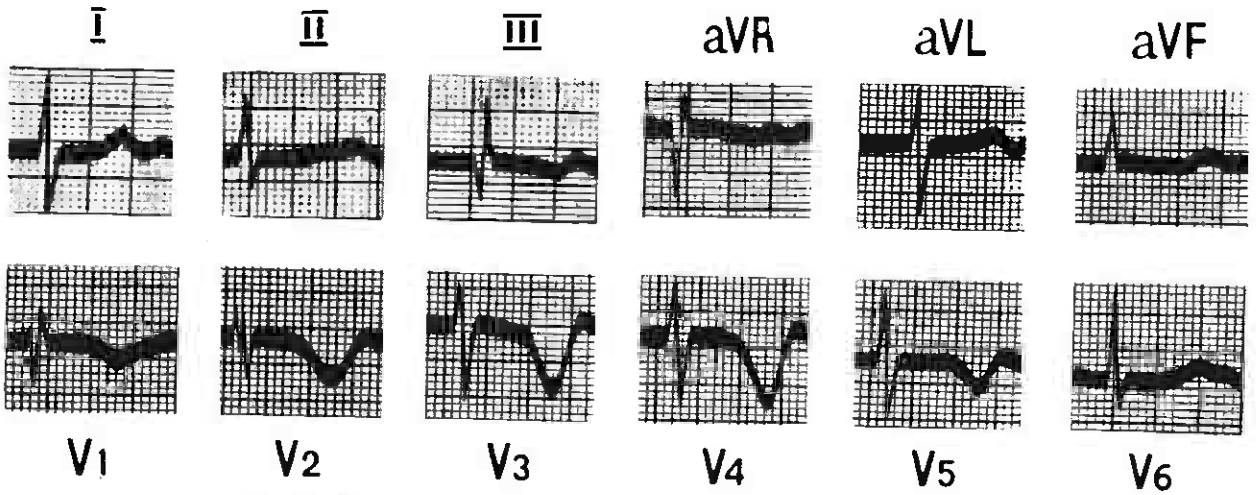

Fig. 4. Case IV. Electrocardiogram showing right bundle branch block and inverted T in leads V_1 - V_5.

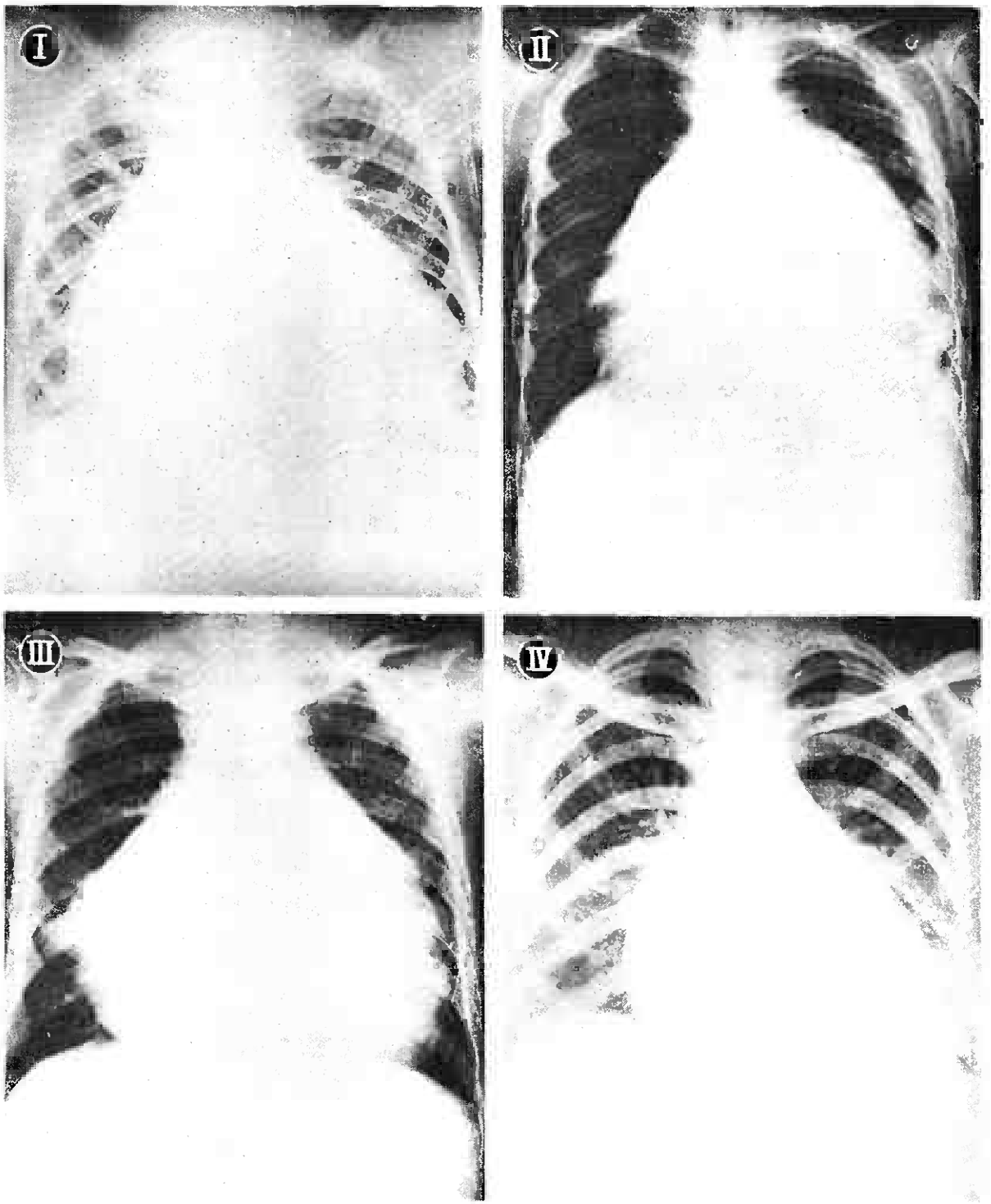

Fig. 5. Posterior-anterior roentgenograms of the chest in Cases I-IV.

of gradual distension of the abdomen for 2½ years. Her feet became swollen intermittently and breathlessness occurred on climbing stairs or after severe exertion. She is under treatment as an outpatient and has had no oedema of the feet on chlortride therapy.

Signs: There were similarities in the physical appearance of these four patients: they were stunted for their ages, their faces were puffy and the abdomens protuberant (Fig. 1). Except for the third patient, the rest had moderate pitting oedema of their feet and legs. There was no clubbing and slight peripheral cyanosis was evident only in the first two patients. The cervical veins were engorged up to the angle of the jaw in all of them, but pulsations were absent except in the last patient in whom a prominent "c" wave was visible. The liver and spleen were much enlarged, felt firm and their edges were palpable 2 to 5 fingerbreadths below the costal margins; only in the last patient was a distinct systolic pulsation of the liver felt. Some free fluid was present in the peritoneal cavity in the first two.

The apex beat was not felt and on percussion the cardiac dullness was found to extend from the left anterior axillary line to beyond the right mid-clavicular line in Cases I to III. Pulsations over the praecordia were absent except in the last patient in whom a distinct systolic pulsation was felt in the left 2nd and 3rd intercostal spaces over an area extending 6 cms. from the left sternal border.

The findings, on auscultation, in the first patient were that of auricular fibrillation; a faint systolic murmur was heard to the left of the lower part of the sternum and in the mitral area. In the second patient, the rhythm was regular with a third heart sound in the mitral area and there were no murmurs; a week before her death, she developed auricular fibrillation. The third patient had auricular fibrillation without murmurs. The cardiac rhythm in the fourth patient was regular and dual; a moderately loud systolic murmur was localized to the mitral area and a faint, short mid-diastolic murmur was present in the left parasternal area in the fourth and fifth intercostal spaces. The heart rate in this patient was slow at 50 per minute.

Electrocardiogram: The findings were that of slow auricular fibrillation in the first three patients (Fig. 2). In the last one, there was complete heart block, the ventricular rate being regular at 50 per minute: the voltage of P was very low and QRS prolonged to 0.12 second with inverted T waves in leads VI to V5 (Figs. 3 and 4).

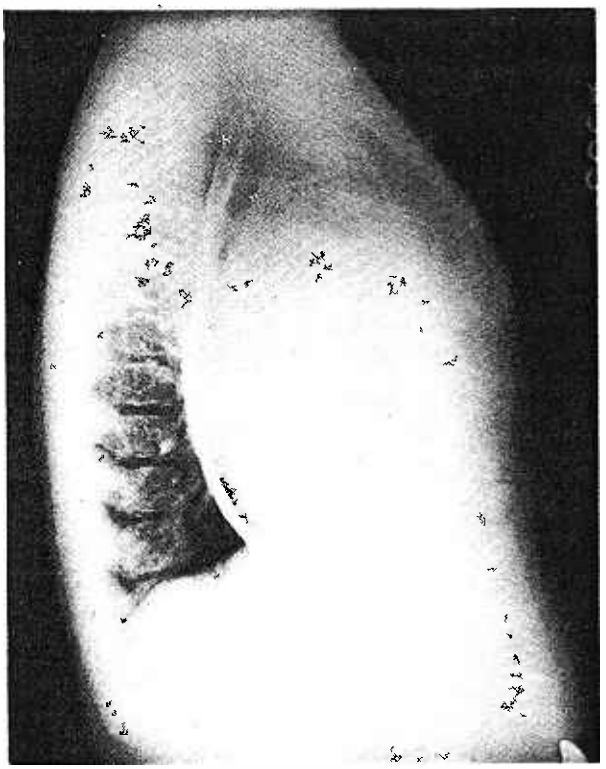

Fig. 6. Case I. Roentgenogram, right oblique view, showing anterior heart border flattened against the chest wall and slight backward displacement of the oesophagus.

Radiologic findings: These were similar in all the four patients so examined (Fig. 5). The heart was grossly enlarged to the right and left with rounded prominence of both borders. The vascular pedicle was relatively small and the main pulmonary artery segment and aortic knob were difficult to identify. The anterior border of the heart was flattened against the chest wall in both oblique views and the oesophagus was displaced slightly backwards by the large heart (Fig. 6). Pulmonary vascular markings were considerably diminished and cardiac pulsations of varying force (weak in Case I, moderate in Case II and forceful in Cases III and IV) were present. There was no pleural effusion. The over-all impression was that of an enlarged heart with a globular configuration and clear lung fields such as is seen in massive pericardial effusion.

Angiocardiography: This investigation, made in Case IV, demonstrated gross dilatation of the right atrium (Fig. 7) and considerable delay in emptying of this chamber so that contrast material remained visible in the last roentgenogram taken 14 seconds from the start of the injection. There appeared to be a notch on the inferior border far over towards the left demarcating the "atrialised" portion of the

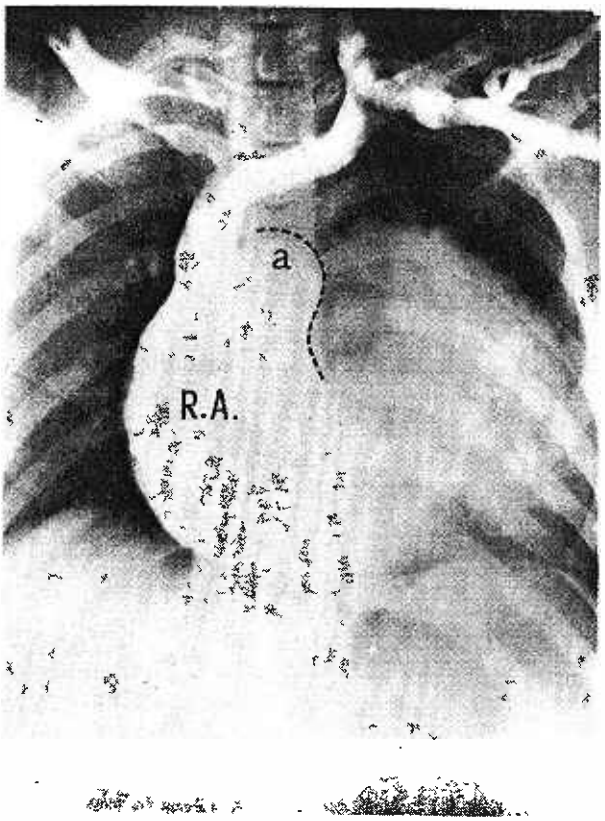

Fig. 7. Angiocardiogram immediately after intravenous injection showing large right atrium (R.A.) and atrial appendix (a). The "atrialized" portion and outflow tract of the right ventricle are becoming visible.

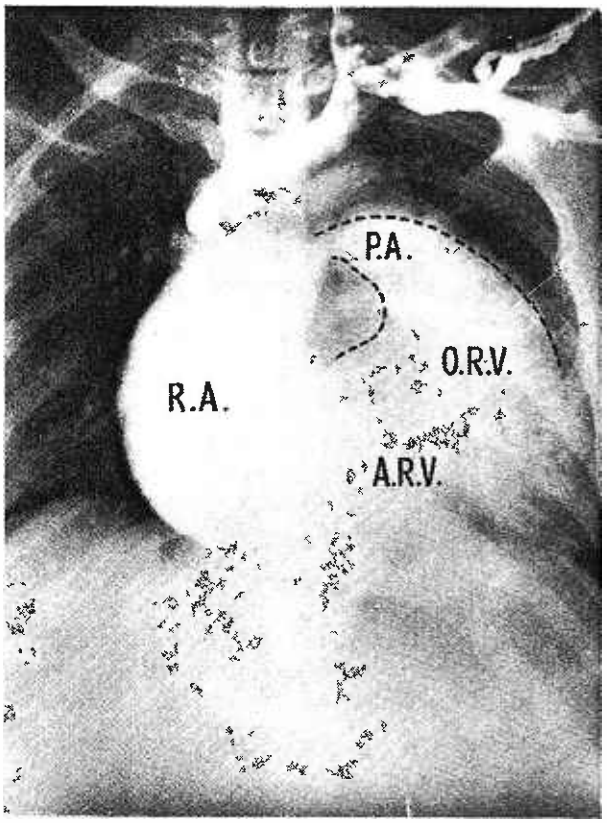

Fig. 8. Angiocardiogram taken 7 seconds from the start of the injection now shows the "atrialized" portion (A.R.V.) and the outflow tract (O.R.V.) of the right ventricle. The pulmonary artery and the right branch are visible.

TABLE I. MAIN FEATURES IN FOUR PATIENTS WITH EBSTEIN'S MALFORMATION

Case	Age	Sex	Symptoms	Cyanosis	Clubbing	Cardiomegaly	Hepato-splenomegaly	E.C.G.	Tricuspid incompetence	Termination
I	14	M	Dyspnoea Abdominal distension Oedema feet 4 years	Slight, peripheral	None	Gross	Present	Auricular fibrillation	Absent	Died after thoracotomy
II	14	F	Cough Dyspnoea Oedema legs 1½ years	Slight, peripheral	None	Gross	Present	Auricular fibrillation	Absent	Died suddenly
III	14	M	None	None	None	Gross	Present	Auricular fibrillation	Absent	Alive
IV	16	F	Dyspnoea Abdominal distension Oedema feet 2½ years	None	None	Moderate	Present	Complete heart block	Present	Alive

right ventricle from the outflow tract (Fig. 8). The pulmonary vessels were poorly filled and opacification of the left chambers of the heart and aorta was not present. There was no interatrial shunt.

NECROPSY REPORT

Necropsies were performed on Cases I and II and on two other patients who had died suddenly and unexpectedly in their homes. One was a 14-year-old Chinese girl (Case V) who was apparently quite well when one morning her brother heard a thud in the bathroom and went in to find her unconscious over the clothes she was washing; she died before she could be brought to hospital. Careful interrogation of the relatives revealed that at no time had she complained of breathlessness or of swelling of

the abdomen and legs. The other patient (Case VI) was a male Indian, aged 17 years, who suddenly collapsed and died; the only relevant history obtainable was that for some months he had complained of anorexia and general weakness.

The pathologic features in all these four cases were remarkably constant. On opening the thorax and abdomen, the heart was seen to occupy more than three-fourths of the thoracic cavity and the lower borders of both liver and spleen extended well below the costal margins (Fig. 10). The liver was finely cirrhotic in Cases I, II, and V, and coarsely nodular in Case VI (Fig. 21); the spleen and kidneys were congested. In Case I, the pericardial sac contained about 150 ml. of bloody fluid and was thickened and adherent in places. The pericardium was normal in the remaining three, but the sac was grossly distended with clear yellow fluid, 500 ml. in Case II, 1200 ml. in Case V, and 300 ml. in Case VI.

The gross enlargement of the heart in every instance was due to an enormously dilated, thin-walled right atrium which formed more than one-half of the anterior surface of the heart. The left ventricle occupied very little of this surface and none of the apical portion; in Case V the left ventricle was completely displaced behind the right ventricle (Fig. 12). The superior and inferior vena cavae opened normally into the right atrium, but their orifices and that of the coronary sinus were widely dilated. The atrial and ventricular septa were intact in every case. The abnormalities were confined to the right atrium and ventricle, the rest of the heart being normal.

Case I: The internal circumference of the right atrium measured 18 cms. Over the area of the right auricular appendix, there was an adherent oval thrombus measuring 11 by 7 cms. (Fig. 9). The tricuspid valve was represented by rudimentary cusps whose line of attachment

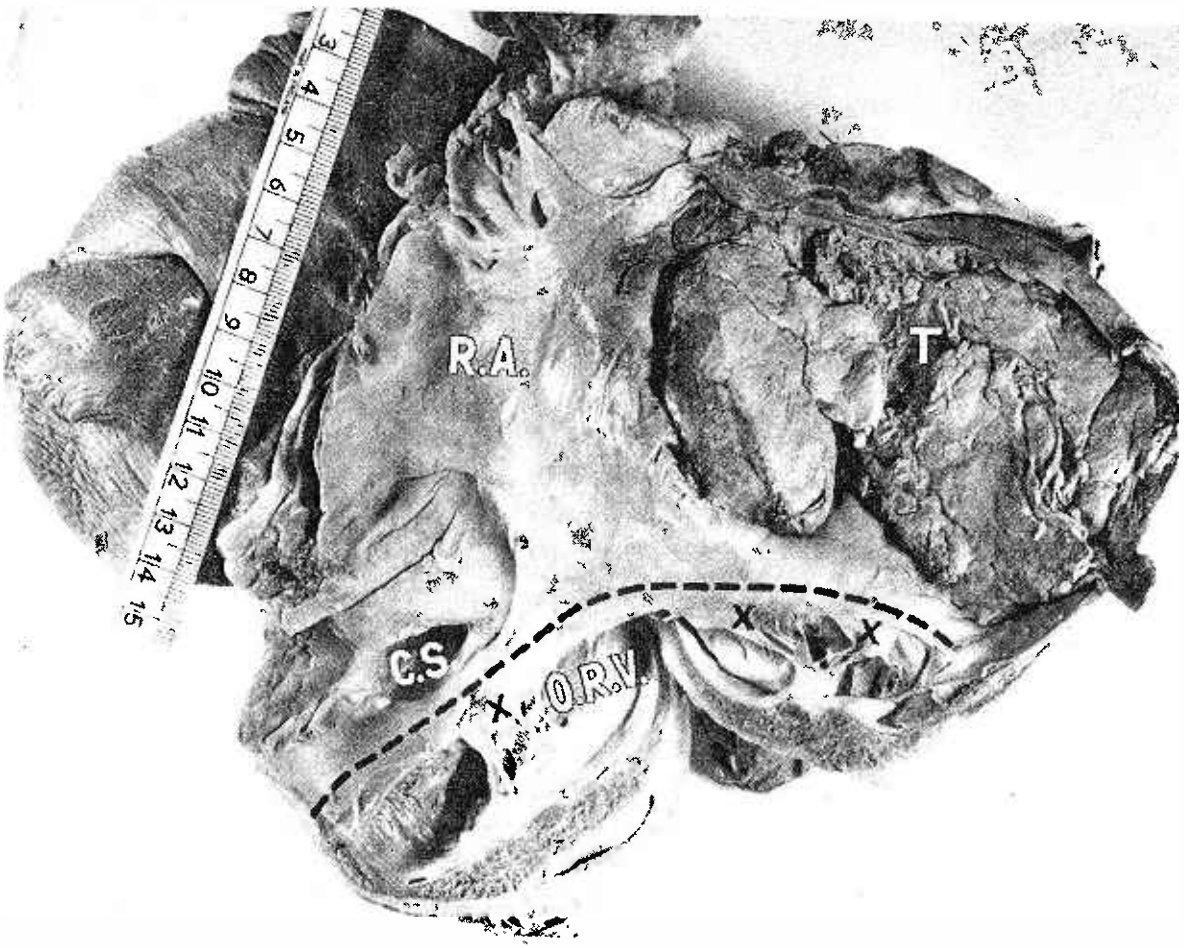

Fig. 9. Case 1. The interior of the right side of the heart was exposed by making a cut along the right cardiac border. The broken line indicates the actual position of the tricuspid valve ring from which the rudimentary cusps (X) arise. R.A. = right atrium. C.S. = large orifice of coronary sinus. T = thrombus. O.R.V. = outflow tract of right ventricle.

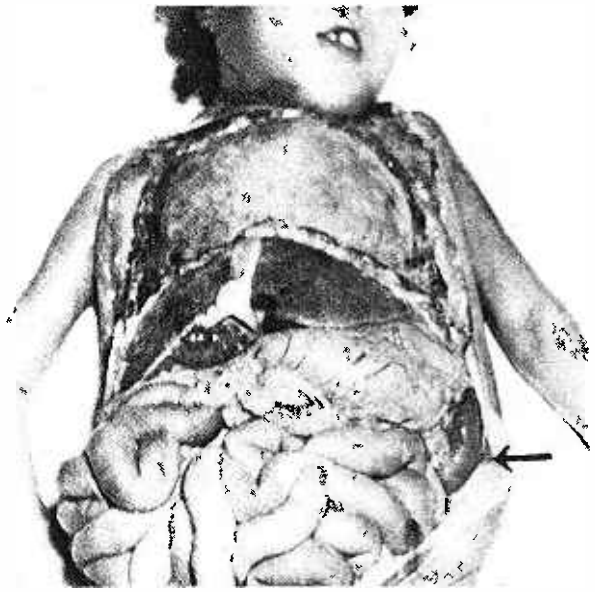

Fig. 10. Case II. Viscera in situ. The heart fills the thoracic cavity and the liver and spleen (arrow) extend well below the costal margins. The pericardial sac contained 500 ml. of fluid.

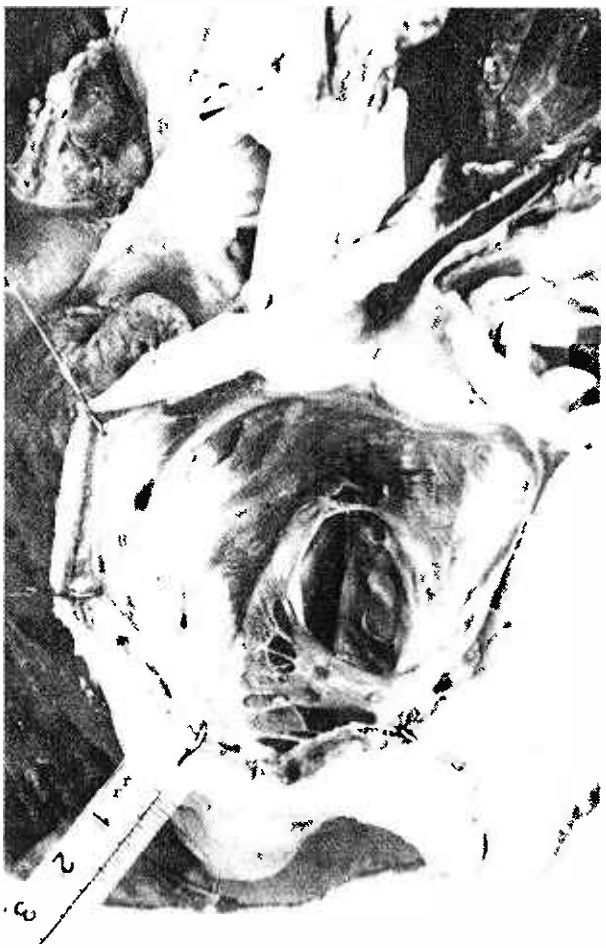

Fig. 11. Case II. Small right ventricle with dilated tricuspid ring and anterior cusp; the posterior and medial cusps were rudimentary and are not seen in the picture.

was displaced downwards into the right ventricle thus reducing the capacity of this chamber. The atrioventricular opening measured 15 cms. in circumference and the valve was obviously incompetent. The diminutive anterior cusp was attached by shortened thin chordae tendinae to a small flat papillary muscle. The posterior and medial cusps were fused to form a single small membranous flap part of which merged into the endocardium while the remaining portion was inserted by short thin chordae tendinae into the ventricular wall. The right ventricular chamber was small consisting mainly of the outflow tract; the wall was not hypertrophied. The endocardium was normal except for an area immediately below the tricuspid valve where it was much thickened and opaque. Liver: 1,785 grams. Spleen: 190 grams.

Case II: The right atrium was grossly dilated and showed an adherent thrombus in the auricular appendage. The tricuspid ring was situated lower than normal and dilated to a circumference of 12 cms. The anterior cusp was narrow and attached by short chordae tendinae to a normal papillary muscle; the posterior and medial cusps were rudimentary membranes attached by a few thin short chordae tendinae to vestigial papillary muscles represented by small protruberances on the endocardial surface. The functioning right ventricle, which was diminished in size and showed no hypertrophy, consisted only of the outflow tract (Figs. 10 and 11). The endocardium in the region of the tricuspid valve and below it was thickened and opaque. Liver: 1,210 grams. Spleen: 300 grams.

Case V. The right atrial cavity was enormous, about 15 cms. in diameter, and contained a large thrombus (6 x 4 cms.) which was adherent to the auricular appendage. The tricuspid ring was widely dilated, measuring 14 cms. in circumference. The valve cusps were very poorly developed and could just be identified. The anterior one was attached by thin short chordae tendonae to a small papillary muscle as was the posterior one; the medial cusp was a small thin membrane attached directly to the endocardium. Due to the low origin of the tricuspid valve, the right ventricular cavity was reduced in size and consisted for the most part of the outflow tract of this chamber. The ventricular wall was not hypertrophied, but the endocardium was thickened and opaque except over the infundibular area (Figs. 12, 13 and 14). Liver: 1,150 grams.

Case VI: The right atrium was grossly dilated and there were thrombi in the auricular

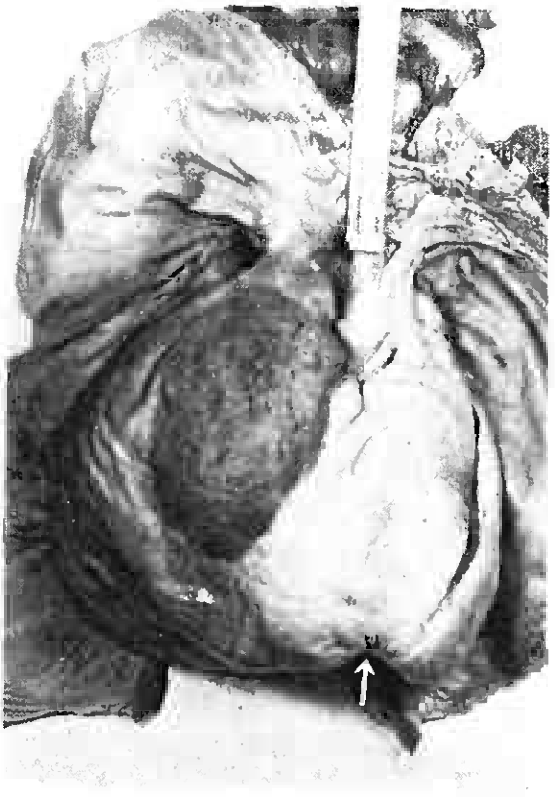

Fig. 12. Case V. Anterior surface of the heart consisting of right atrium and right ventricle only. The site of the tricuspid ring is indicated by the arrow. A cut has been made in the anterior wall of the right ventricle.

Fig. 13. Case V. Small right ventricle consisting of the outflow tract. The dilated tricuspid ring together with a rudimentary cusp (arrow) is seen.

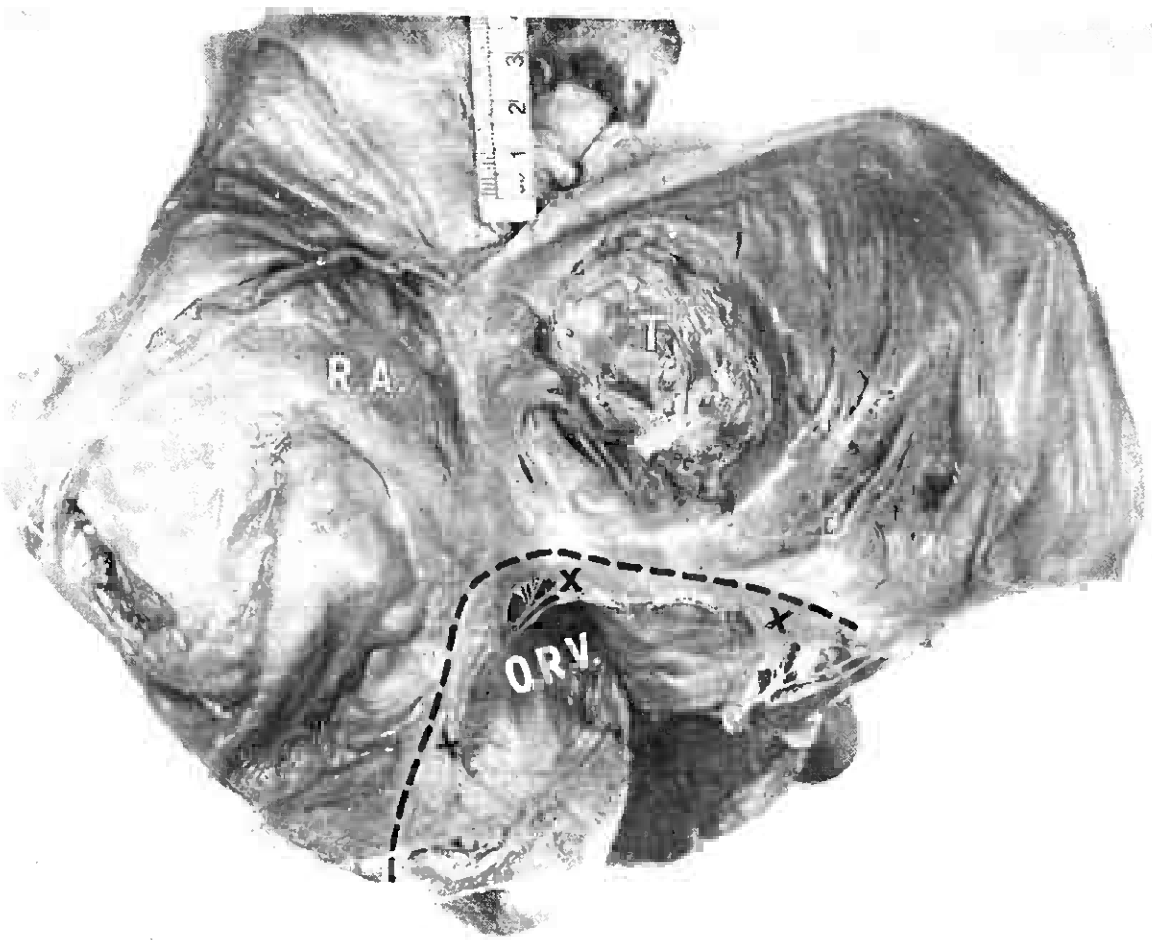

Fig. 14. Case V. The interior of the right side of the heart was exposed by making a cut along the right cardiac border. The broken line indicates the actual position of the tricuspid valve ring from which the rudimentary cusps (X) arise. R.A. = right atrium, T = thrombus. O.R.V. = outflow tract of right ventricle.

appendix. The tricuspid valve was found to be displaced, arising from a lower position than normal and the atrioventricular opening was dilated to 12.5 cms. in circumference. The

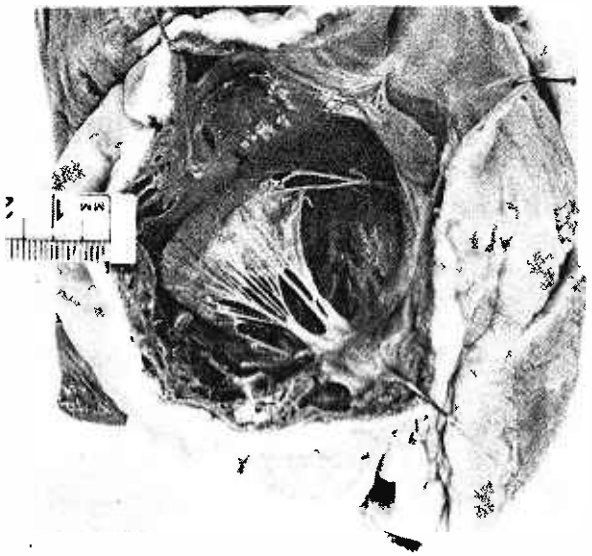

Fig. 15. Case VI. Small right ventricle, dilated tricuspid ring and anterior cusp with diminutive papillary muscle. The posterior and medial cusps were rudimentary and are not seen in the picture.

anterior cusp appeared normal in size, but it was attached by chordae tendinae to a diminutive papillary muscle (Fig. 15). The posterior cusp was very poorly developed, consisting of a narrow thin membrane with short chordae tendinae inserted into a tiny papillary muscle. The medial cusp was represented by a narrow thin membrane which had fused with the endocardium. The right ventricle was reduced in size, but was not hypertrophied. The endocardium in the region of the tricuspid ring and below it was thickened and opaque. Liver: 1,535 grams. Spleen: 240 grams.

Histologic picture: This was similar in all four cases. The tricuspid valve leaflets were represented by short fibrous tags continuous with the endocardium. Immediately above the valve, the wall of the atrium was very thin and represented the "atrialized" portion of the right ventricle (Fig. 16). The myocardium of the right side of the heart showed the greatest degree of abnormality in the atrium where degeneration to a varying extent represented by vacuolation and separation of fibres was noted. Small areas of haemorrhage and necrosis were seen in all four cases and calcification of necrotic muscle was present in Case II. A striking feature was the large size, the hyperchromatism and the abnormal shape of many of the muscle nuclei which were found scattered evenly throughout the entire sections of atrium studied; these changes were present to a lesser degree and extent in the adjacent part of the ventricle. The nuclear changes were not always found in areas of degeneration; in many instances they were seen in fibres where the cross striations were present (Figs. 17 and 18). Fresh

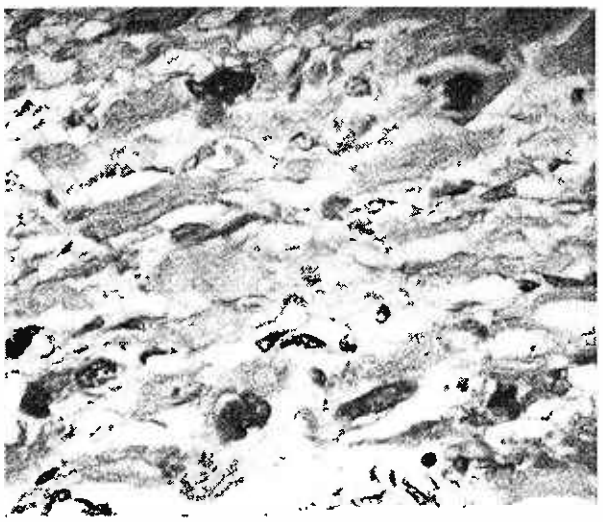

Fig. 17. Section of myocardium showing large muscle nuclei and degenerate fibres. Haematoxylin and eosin stain X 500.

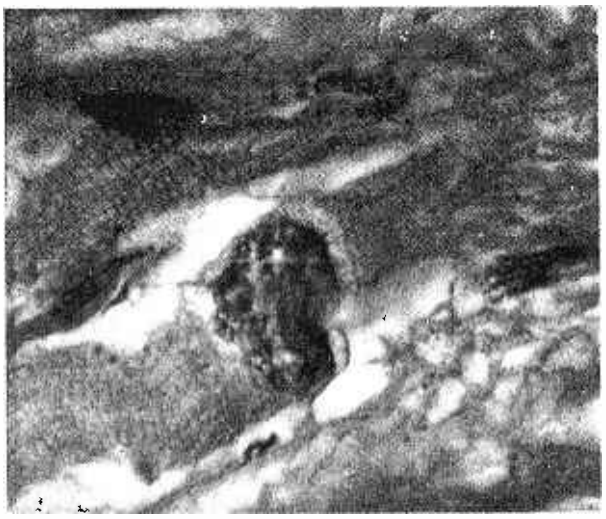

Fig. 18. Giant myocardial nuclei. Note presence of cross striations. Haematoxylin and eosin stain X 1180.

and organized thrombi were attached to the right atrial endocardium and there were focal areas of lymphocytic infiltration in the pericardium over the atrium. Fibroelastic thickening of the endocardium of the right ventricle with extensions between deeper muscle bundles or around the sinusoids was present in all four cases (Fig. 19). No occlusion was seen in the

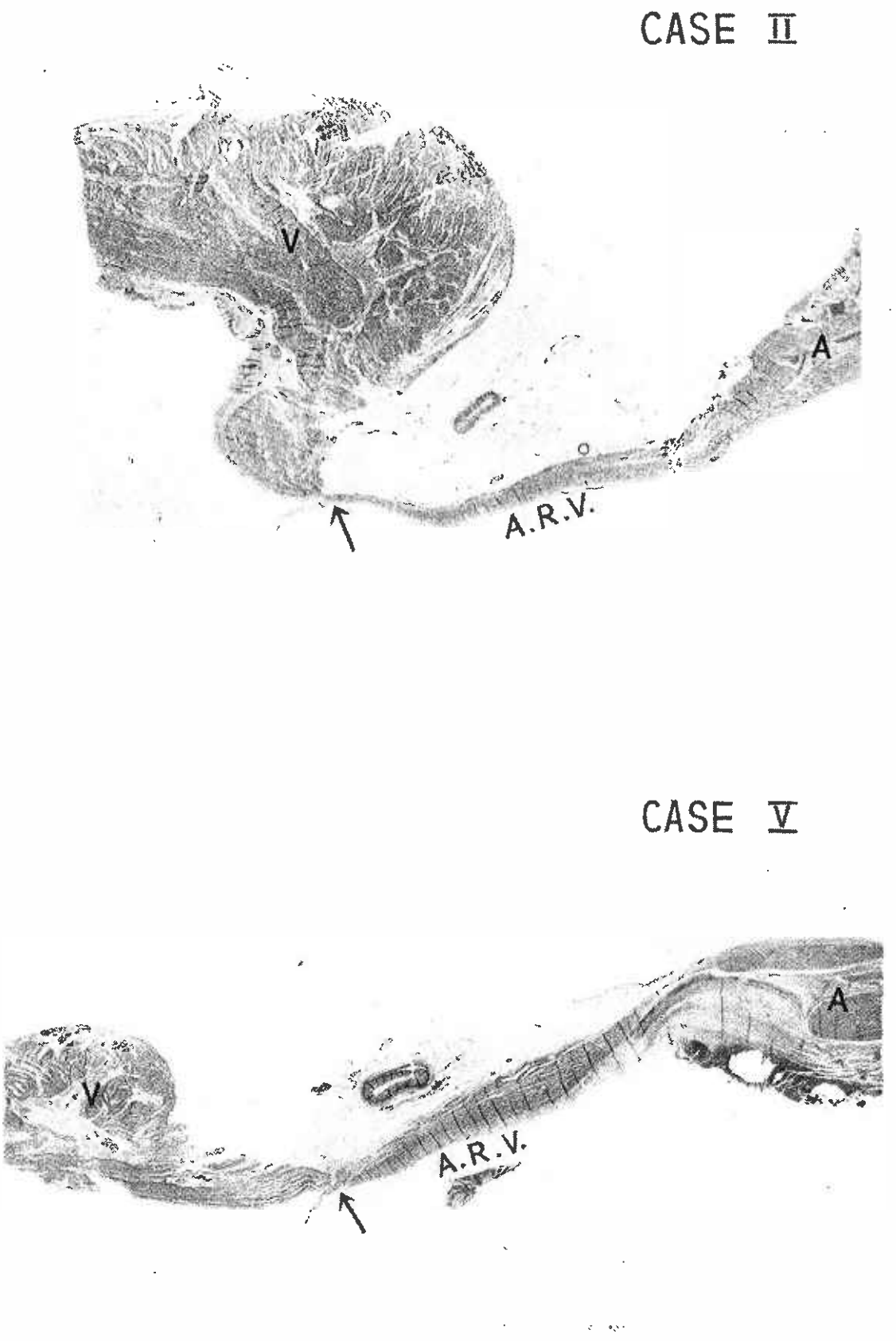

Fig. 16. Section through tricuspid valve ring and adjoining atrium (A) and ventricle (V). Arrow indicates point of attachment of rudimentary cusp. Note the extreme thinness of the "atrialized" portion of the right ventricular wall (A.R.V.). Haematoxylin and eosin stain X 4.5.

right coronary artery, but in all four cases this vessel in cross section showed fibroelastic thickening of the intima with fragmentation of the internal elastic lamina. In Case II a branch of this main vessel, while in the pericardium, showed calcification of the medial coat with intimal proliferation almost obliterating the lumen (Fig. 20).

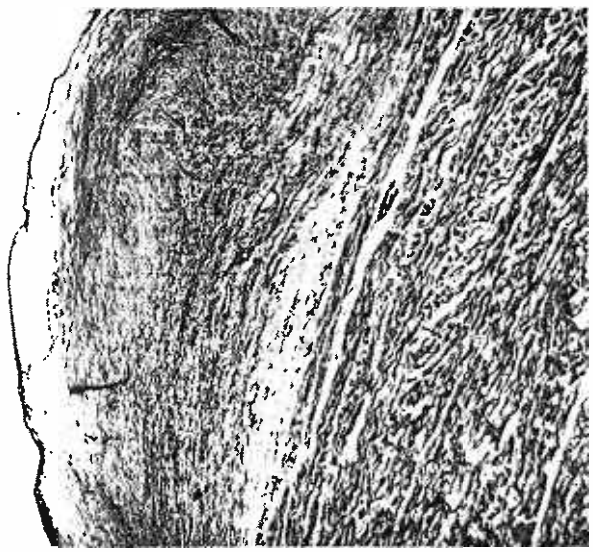

Fig. 19. Section of myocardium showing fibroelastic thickening of endocardium of right ventricle. Verhoeff-van Gieson stain X 45.

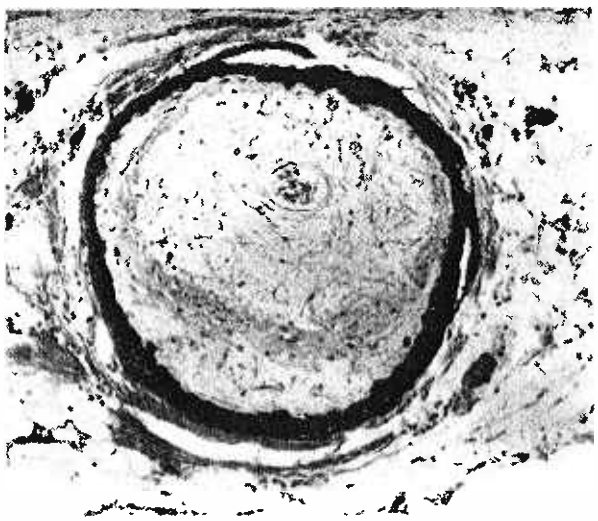

Fig. 20. Section showing medial calcification of coronary artery with intimal hyperplasia. Haematoxylin and eosin stain X 150.

Microscopic examination of all four livers showed a broadly similar histologic appearance. Many centrilobular areas were replaced by degenerate liver cells, collapsed reticulin framework, young fibrous tissue, capillaries and in some places lymphocytes. The vascular fibrous tissue extended in radiating bands to adjacent portal tracts and surrounding centrilobular zones. This produced a pattern of irregular nodules of liver cells many of which showed regenerative hyperplasia (Figs. 22 and 23). Loss of liver tissue, fibrosis and resulting distortion of architecture varied greatly from case to case. The central veins were not prominent in the collapsed areas and in some cases were replaced by several dilated spaces which communicated with dilated channels between surrounding cords of liver cells. The walls of these spaces sometimes contained surviving liver cells and elsewhere was composed of collapsed reticulin and fibrous tissue.

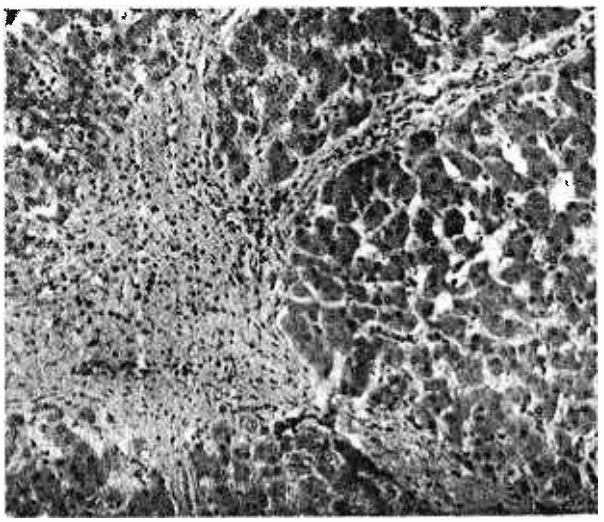

Fig. 22. Section of liver. Centrilobular collapse with radiating bands of fibrous tissue separating nodules of liver cells showing regenerative hyperplasia. Haematoxylin and eosin stain X 150.

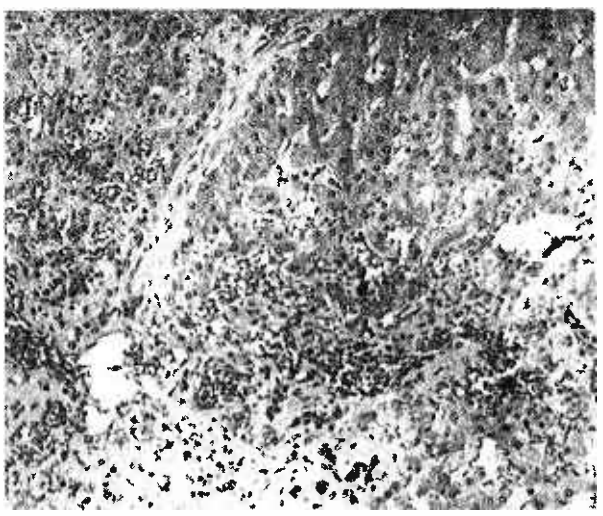

Fig. 23. Section of liver showing centrilobular congestion and collapse with fibrous septum connecting to a portal tract. Haematoxylin and eosin stain X 150.

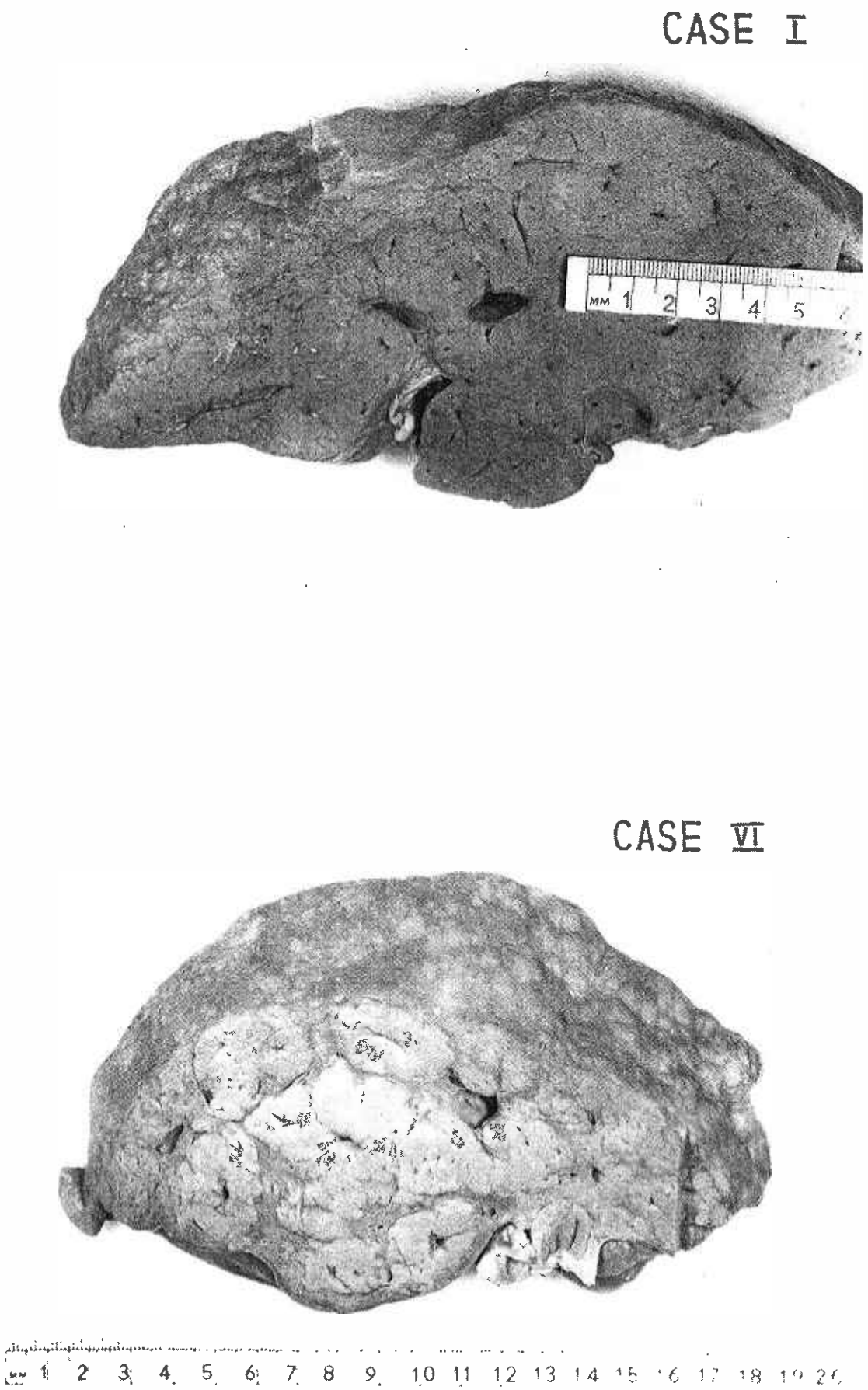

Fig. 21. Cirrhosis of liver.

DISCUSSION

The diagnosis of Ebstein's malformation was made in the first patient only at thoracotomy. In the remaining three patients seen during life, the clinico-roentgenologic picture of a grossly enlarged heart, venous congestion and hepato-splenomegaly was similar to that of the first patient and the same diagnosis was therefore made; this was confirmed at necropsy in Case II and by means of an angiocardiogram in Case IV. Any other diagnosis except that of a massive pericardial effusion, such as is seen in tuberculosis seemed unlikely. There were, however, no toxic manifestations in our patients, they were all ambulant before coming into hospital, the duration of symptoms (where present) extended to some years and there was relatively good exercise tolerance in the presence of considerable cardiomegaly, all of which features would tend to exclude a large pericardial effusion. Nevertheless, an effusion was present in Case I and demonstrated at necropsy in Cases II, V, and VI, so that a mistaken diagnosis of pericardial effusion may be made especially in acyanotic cases if it is not realised that the effusion contributes only partly to the large cardiac shadow. It may be therefore necessary to delineate the heart borders by replacing aspirated fluid by air or by using a catheter to identify the right border of the right atrium.

In the patients in this series there was no central cyanosis, but when it is present other congenital cardiac defects characterized by cyanosis and an inadequate pulmonary blood flow, such as Fallot's tetralogy, pulmonary stenosis with interatrial defect and tricuspid stenosis, must be considered. Confusion is unlikely to arise in the severe cases of Ebstein's malformation where cardiac enlargement is gross. A feature to note is that distinctive praecordial pulsations are not present except sometimes over the outflow tract of the right ventricle in the second and third left intercostal spaces parasternally as was noted in Case IV and as reported by Blacket et al. (1952). Absence of marked pulsation to the left of the sternum is mentioned by Medd et al. (1954) as a useful sign in differentiating Ebstein's malformation from other forms of cyanotic congenital heart disease in which there is right ventricular hypertrophy.

Widely varied **auscultatory findings** have been reported in Ebstein's malformation, but none are sufficiently specific to enable a diagnosis to be made. Of 71 cases reviewed by Kilby et al. (1956), the presence or absence of a cardiac murmur was mentioned in 63. No murmurs were present in 8 of the latter group, and slightly less than half of the remainder had both systolic and diastolic murmurs. In 25 cases only systolic murmurs were present. A superficial diastolic scratch was heard, usually in the 3rd left intercostal space parasternally, in eight out of ten cases examined by Wood (1956) who thought that the sound was more like a diastolic pericardial friction over the distended right atrium than a true intracardiac murmur. The murmurs heard in two of the patients in the series reported in this paper were not considered of diagnostic significance.

Of our four patients seen during life, **hepatic pulsation** together with jugular venous pulsation was present only in Case IV. Ebstein (1866) noted this physical sign in his patient, but subsequent authors have recorded it only infrequently (Taussig, 1947; Gotzsche et al., 1954). In two recent reports of large series of cases (10 by Mayer et al., 1957; and 23 by Schielbler et al., 1959) hepatic pulsation was not found. The lack of clinical signs of tricuspid insufficiency has been repeatedly noted by many authors and various explanations (Kjellberg et al., 1955; Baker et al., 1950) have been given to account for this. From a study of our necropsy cases we are inclined to accept the explanation of Brown et al. (1956) who considered that the absence of signs is due to a very small regurgitant stream from the anatomically small right ventricle entering an abnormally large right atrium which acts as a buffer.

A review of the **electrocardiograms** in 86 cases revealed significantly abnormal findings (Vacca et al., 1958). Right bundle branch block was present in 61 cases, two thirds showed large P waves and one-quarter had findings compatible with a first degree atrio-ventricular block. Abnormalities of rate or rhythm were noted in one quarter of the cases and the Wolff-Parkinson-White syndrome in six instances. According to Sodi-Pollares et al. (1958) the presence of the latter syndrome, type B, in a child's tracing should suggest the possibility of Ebstein's malformation. Paroxysmal supraventricular tachycardia and atrial and ventricular extrasystoles have been reported and, less frequently, atrial flutter (Kistin et al, 1955) and atrial fibrillation (Henderson, 1953; Kilby et al. 1956 Sinha et al., 1960). During cardiac catheterization the increased incidence of arrhythmias which might prove fatal has been emphasized by Engle et al. (1950). The electrocardiogram in three of our patients showed atrial fibrillation; in the fourth there was evidence of complete heart block, an abnormality not previously reported.

The most constant radiologic features in advanced cases consist of a tremendously dilated right heart with normal or decreased pulmonary vasculature and absent left atrial enlargement; pleural effusions are not present (Amplatz et al., 1959). In our patients the enormous globular cardiac shadow with diminished pulmonary vascularity produced a stencilled effect on the skiagram suggestive of a large pericardial effusion, but cardiac pulsations were present thus helping to exclude this diagnosis. However, at necropsy large quantities of fluid varying from 300 ml. to 1,200 ml. were found in the pericardial sac in Cases I, II, V and VI. The effusions were probably produced by increased pressure on the coronary sinuses whose openings into the right atria were found considerably dilated in all these cases. The accumulation of pericardial fluid had probably occurred some time before death contributing to the radiologic picture. Bauer (1945) also reported finding a large pericardial effusion in his case at necropsy, but this finding is not mentioned by other authors and its occurrence seems to be unusual.

The angiocardiographic findings in Case VI are in agreement with those previously reported (Soloff et al., 1951; Blacket et al., 1952; Goodwin et al., 1953; Kistin et al., 1955). The two most characteristic features, an enormous right atrium with considerable delay in emptying and the separation of the functioning portion of the right ventricle from the "atrialized" proximal portion, were present in Case VI; an atrial communication was not demonstrated. The poor filling of the pulmonary vessels and the non-visualization of the aorta were considered to be due to the gross delay of the dye in the right atrium and its considerable dilution before passing into the right ventricle. Soloff et al., (1951) and Kistin et al., (1955) were able to demonstrate a narrow band or notch separating the functioning right ventricle from the auricular portion of the right ventricle. The angiocardiographic picture would appear to be pathognomonic, but Goodwin et al. (1953) believe that extreme right atrial enlargement due to severe pulmonary stenosis may simulate it by displacing the normal tricuspid notch to the left. However, details of the tricuspid region are difficult to identify in cases with a right to left interatrial shunt where the left chambers of the heart are depicted simultaneously with the right. Kjellberg et al. (1955) consider that the information provided by angiocardiography is of extremely limited value and maintain it is dangerous to carry out this investigation. In the only reported fatality due presumably to angiocardiography, the patient developed marked bradycardia and coma during the procedure and died 36 hours later (Mayer et al., 1957).

Sudden death, for which no cause other than the cardiac malformation could be determined, has been recorded fairly frequently (Bauer, 1945; Engle et al., 1950; Yater and Shapiro, 1937; Kerwin, 1955). Cardiac arrythmias may have been supervened. In four of the patients in this series, the termination was sudden and unexpected in three. Other reported causes of death have been brain abscess (Barger et al. 1951), paradoxic embolism (Walton and Spencer, 1948) and pulmonary tuberculosis (Ebstein, 1866). Congestive heart failure has been noted in approximately one-third of the recorded deaths (Kilby et al. 1956). Although there have been survivals even up to the age of 79 years (Adams and Hudson, 1956), the average age at the time of death has been 23 years (Vacca et al., 1958). Three of our patients were aged 14 years and one 17 when they died.

Surgical treatment has been attempted in this congenital malformation for the closure of an associated atrial defect, to eliminate the veno-arterial shunt and the possibility of paradoxic emboli and cerebral abscesses. Of four patients who were thus operated on, one was greatly improved but the others died during induction of anaesthesia or soon after operation (Kilby et al. 1956). Recently, a new surgical procedure has been suggested by Hunter and Lillehei (1958) in which the displaced leaflets are brought up to the true annulus by means of a plicating technique that excludes the thin wall of the "atrialized" portion of the right ventricle. Fenestrations in the leaflets of the tricuspid valve are repaired by fine stitches and any patent foramen ovale or atrial septal defect is closed. Two poor-risk patients on whom this surgical repair was attempted did not survive (Schiebler et al. 1959). Another surgical procedure proposed is that of anastomosis of the superior vena cava to the pulmonary artery (Sanger et al., 1959). From a study of our cases, this shunt operation appears to be a more feasible approach to the treatment of Ebstein's malformation. It would result in relief of the marked congestion of the systemic venous system and right atrium and also increase the blood flow to the lungs.

The necropsy findings in our four cases were characteristic of Ebstein's malformation. The primary abnormality lay in the development of the tricuspid valve, the papillary muscles and the chordae tendinae, all of which were rudimentary and could not have functioned effectively enough to prevent regurgitation through the dilated atrio-ventricular opening. The size of the functioning right ventricle was greatly diminished and limited to the outflow portion by the low origin of the tricuspid valve, while the right atrium which included the proximal or "atrialized" portion of the right ventricle was greatly dilated. Large thrombi were found in each right atrium. An unusual feature was the absence of a patent foramen ovale or an atrial septal defect. Vacca et al. (1958) in reviewing the literature found 57 necropsy cases of which 46 had defects of the septum. The mechanism for a right-to-left shunt was therefore present in a large number, thus accounting for the cyanosis so frequently reported.

Histologic abnormalities are infrequently mentioned. Thickened endocardial plaques noted in a few of the 16 cases reviewed by Yater and Shapiro (1937) were considered by them to be only incidental; in their own case no such change was found. Baker et al. (1950) noted in their first case that sections of the right ventricle showed a normal myocardium, but a thickened endocardium, while Lev et al. (1955) found fibroelastosis of the proximal right ventricular chamber. In each of our four cases the endocardium of the functioning right ventricular chamber showed fibroelastic thickening. The unusual histologic appearances of the myocardial nuclei and the intimal changes in the coronary artery have not been recorded previously and their pathogenesis is obscure.

In all our four cases that came to necropsy, the wall of the right ventricle proximal to the abnormal tricuspid valve was very thin whereas the ventricular wall of the distal or outflow chamber was of normal thickness. Engle et al. (1950) mention that thinness of the upper portion of the right ventricle has been noted in nearly all the recorded cases including their own and consider this feature an integral part of Ebstein's malformation. They postulated that both the maldevelopment of the proximal right ventricle and tricuspid valve resulted from a defect of the visceral coelomic wall destined to give rise to the right ventricular myocardium. However, Edwards (1953) pointed out that one of his cases failed to show this feature, perhaps because the patient was only two months old. He suggested that the thinness of the right ventricular wall may be the result of the altered dynamics secondary to the deformity of the valve rather than a primary abiotrophy of the muscle tissue. Kerwin (1955) noted in his patient, who was 18 years when he died, that the wall of the proximal portion of the right ventricle was thicker than normal, and, therefore, felt that a visceral coelomic defect is not an integral part of this malformation. He presumed that an unusual degree of tricuspid stenosis could account for the proximal hypertrophy; in his patient the tricuspid valve was a membranous sheet encircling the atrio-ventricular orifice and extended as a drape from the A-V opening almost to the apex of the right ventricle. No specific histologic defect has been noted in the myocardium of the proximal right ventricle (Engle et al., 1950; Mayer et al., 1957).

A distinctive and unusual feature in the present series was the marked hepato-splenomegaly with cirrhotic changes in the liver not mentioned by other workers. In the extensive review of Ebstein's malformation by Vacca et al. (1958) no mention is made of this finding. In our cases the histologic changes in the liver were marked and consisted of areas showing collapsed reticulin fibres with surrounding areas of regeneration. Some of these collapsed areas did not show fibrosis or deposition of collagen, in other areas this was present to a small extent and in others dense fibrosis or proliferating fibrous tissue with or without inflammatory cells were seen. Long standing passive congestion of the liver had proceeded to the stage of irregular fibrosis with regenerative hyperplasia as is seen in cirrhosis. Popper and Zak (1958) have emphasized that only in very severe and long-standing cardiac failure as in tricuspid insufficiency and constrictive pericarditis do the pathologic changes in passive congestion become extensive enough to justify the term cardiac cirrhosis. A reference to histologic changes in the liver is made by Lev et al. (1955) who found marked chronic passive hyperaemia with central necrosis. Of eight necropsy cases, chronic passive congestion often with atrophy of the surrounding liver cells was usually noted; in one case, the changes had proceeded to cardiac cirrhosis (Schiebler et al., 1959). We consider our patients who came to necropsy to be severe examples of Ebstein's malformation with marked venous congestion of long duration to produce the gross changes mentioned above. Unlike many of the cases recorded in literature, there was no defect in the atrial septum so that the pressure in the right atrium

was not relieved by a venous-arterial shunt. This probably accounted, in part, for the severe congestive changes noted in these patients.

SUMMARY

Six cases of Ebstein's malformation are reported. The notable clinical features seen in four patients were gross enlargement of the heart, venous congestion and marked hepatosplenomegaly. There was complete heart block in one patient and auricular fibrillation in the remaining three.

The unusual pathologic features in the four cases that came to necropsy were the large pericardial effusions associated with the cardiomegaly, the absence of an atrial septal defect or patent foramen ovale, the gross enlargement of liver with cirrhotic changes and the large spleen. Histologic examination of heart muscle revealed certain unusual features which are described.

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