Sudden Death in Idiopathic (Congenital) Right Atrial Enlargement

Postmortem Findings and Review of Cases

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SUMMARY

A case of idiopathic enlargement of the right atrium is presented and 13 other cases are reviewed. The patient whose case is reported died suddenly at home 6 months after onset of atrial flutter. This represents the first case in which postmortem findings are available. The patient demonstrates that the anomaly cannot be considered entirely benign as has been suggested in previous reports. Other complications that may occur particularly in cases of massive atrial enlargement are atrial arrhythmias, syncope, and right ventricular compression. The importance of recognizing this condition as a clinical entity is stressed because surgical excision of a portion of the enlarged atrium appears to be both easy and curative and because these cases may be mistakenly diagnosed as mediastinal tumor or pericardial effusion. The possible relationship of this apparently congenital abnormality to Uhl's anomaly is discussed.

Additional Indexing Words:
Right ventricular compression  Supraventricular arrhythmia  Pericardial effusion
Uhl's anomaly  Parchment heart

In 1961, Pastor and Forte' first drew attention to a clinical entity which they termed "idiopathic enlargement of the right atrium." They reported three cases. In 1962 Saigusa and associates reported one case; in 1965 Sumner and co-workers described four cases, and in 1969 Sheldon and associates reported three additional cases plus one with a right atrial diverticulum. The clinical picture in the 11 patients with idiopathic right atrial enlargement was similar. In each instance, attention was first focused on the heart by the unexpected finding of cardiac enlargement on routine x-ray examination in an otherwise asymptomatic patient. The clinical findings were normal; murmurs, when present, were nonspecific grade I/IV systolic murmurs, and the ECG was normal in all but two patients. The ECG abnormalities encountered were atrial premature beats, coronary sinus rhythm, and aberrant ventricular conduction. Cardiac catheterization was carried out in six patients with angiography in five of these, and an enlarged right atrium was the only abnormal finding in all six. The diagnosis in each instance was made on clinical grounds and was confirmed in two cases at surgery. Postmortem findings have, however, not been described to date, and indeed, previous reports have stressed the benign nature of the condition. We have recently encountered a...
patient with idiopathic right atrial enlarge-
ment and cardiac arrhythmias who died
suddenly. This is the first case in which the
postmortem findings are reported.

Report of Case

A 16-year-old boy was admitted to the hospital
with a history of exertional dyspnea and
palpitations of 1½ months’ duration. He was an
ardent athlete and had participated actively for
many years in competitive sports, such as football,
discus throwing, and track. One and one half
months prior to admission, while on a 3-mile
walk, palpitations, shortness of breath, and
faintness began suddenly and required him to
stop and rest. Thereafter, these symptoms
recurred with exertion, especially on running or
climbing. Since these symptoms reduced his
athletic performance, he finally consulted his
physician. A routine chest x-ray taken as part of
the examination showed a massive cardiac
silhouette (fig. 1), and the patient was admitted
to the hospital. No additional history could be
elicited except that another child in the family
had had influenza 1 month previously. The boy
had been adopted as a baby and no family history
was available.

On examination the boy was of a healthy
athletic build and in no distress. The pulse varied
in rate from 70 to 120 with runs of irregular
beats. The heart was quiet to palpation. The first
and second heart sounds were distant. No other
heart sounds or murmurs were heard, but a sys-
tolic pericardial friction rub was heard over the
lower left sternal border. The lungs were clear,
and there was no venous engorgement or hepatic
enlargement. Blood pressure was 120/80 mm Hg,
decreasing by 15 to 20 mm on deep inspiration.
The right leg was ½ inch shorter than the left, the
medial malleolus being unusually prominent. The
rest of the examination was unremarkable.

ECG (Fig. 2)

The ECG showed atrial flutter with a 2:1
block, alternating with a 3:1 block, and occasion-
al ventricular premature contractions. QRS vol-
tage was decreased in all leads.

Chest Roentgenogram

The large cardiac silhouette (fig. 1) was
thought to be due to a large mass situated within
the pericardium or to pericardial effusion. There
was no compression or displacement of the
trachea, esophagus, or aorta. Fluoroscopy demon-
strated pulsation of the superior and right border
of the mass and a definite change in shape of the
mass with position of the patient. Hilar markings
appeared normal.

In view of the recent onset of symptoms, an
enlarged heart, distant heart sounds, pulsus
paradoexus, pericardial friction rub, and low voltage
ECG, it was felt that pericardial effusion with
imminent cardiac tamponade was the most likely
diagnosis. Consequently, pericardial tap was
carried out under direct vision, using an image
intensifier. The puncture needle penetrated a
thin, dilated myocardium, and venous blood was
withdrawn. No pericardial effusion was found.

Because of the atrial flutter and the cardio-
megaly, digitalization was instituted prior to
cardiac catheterization. In the meantime the
patient recalled having had a chest x-ray at a
mobile T. B. unit at the age of 7 years. These
films were sent for and showed that the same
massive cardiomegaly had been present at that
time, 9 years before the present examination.

Cardiac Catheterization

A no. 7 catheter was introduced via the right
arm and entered the large right atrium in which it
coiled up repeatedly. The coiled catheter showed
that the atrium extended well beyond the left
sternal border; retrospectively it became evident
that the right atrium had been entered during the
preceding pericardial tap. Although repeated
attempts were made to insert the catheter through
the tricuspid valve, these failed. A prolonged or
more vigorous search was not attempted because
of the danger of perforating the huge thin-walled
atrium. There was no evidence of a left-to-right
shunt at the atrial level. The atrial pressure

Figure 1

Chest roentgenogram of patient at age 16 years, 6
months prior to sudden death.

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tracing showed regular a waves at a rate of 350, which, when followed by a QRS complex, were of normal height (4 mm) but rose to varying heights, maximally 12 mm, when contracting against a closed tricuspid valve.

Diatrizoate sodium and meglumine (Renovist), when injected into the right atrium, floated around in the large venous reservoir and slowly disappeared without adequately outlining the right ventricle. When the dose of Renovist was doubled (80 cc), a thin stream of contrast material was drawn rapidly through the extreme left edge of the tricuspid valve and into the right ventricular outflow tract, which appeared to be normal. The right ventricle was never completely outlined. There was no evidence of tricuspid regurgitation. Retrograde catheterization of the left ventricle via the brachial artery demonstrated normal pressures in the left ventricle and aorta and no evidence of a right-to-left shunt. An angiocardiogram revealed an anatomically normal left ventricle and aorta. On comparison of the left and right angiograms, it appeared likely that the right ventricle was of normal size. The cardiac index at rest was abnormally low, 2.35 L/min/m².

In summary, this patient appeared to have an enlarged right atrium with no other cardiac abnormalities and the diagnosis was thought to be idiopathic dilatation of the right atrium or a form of Ebstein's disease.

The patient refused to consider cardiac surgery and was only with difficulty persuaded to give up competitive sports. He was sent home on digitalis because of persistent flutter. On follow-up visits, 2 weeks and 3 months after discharge there was some definite improvement in symptoms corresponding with his reduction of activities. However, atrial flutter persisted, and administration of digitalis was continued. The patient died suddenly at home 5 months after admission to the hospital and 6% months after the onset of symptoms. A routine postmortem examination was refused, but permission was given for the intact heart to be removed from the body by the mortician and the specimen was sent to the Children's Orthopedic Hospital for examination.

Postmortem Findings

Currently at our institution, postmortem hearts with a known or suspected abnormality are being formalin-fixed under controlled pressure as first described by Glagov and associates⁶ and subsequently modified by us. Following fixation, the internal cardiac structures are dusted with barium powder followed by x-ray filming as originally described by Wright and co-workers.⁶ The

*Electrocardiogram on admission shows atrial flutter with occasional ventricular premature beats and an unexpectedly low QRS voltage for an athletic 16-year-old youth.*

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Heart after controlled pressure fixation. More than half of the anterior aspect of the specimen is comprised of right atrium. Everything to the left of the prominent horizontal atrioventricular sulcus represents right atrium.

The patient's heart, freed of blood, weighed 570 g. The appearance of the heart after controlled pressure fixation is shown in figure 3. More than half of the anterior aspect of the heart consisted of right atrium, which accounted for approximately two thirds of the total volume of the specimen. The anterior atrioventricular sulcus was unusually deep between the right atrium and the right ventricle, and the atrium overlapped the base of the right ventricle. The cardiac apex was directed toward the left but was blunted, and composed entirely of left ventricle. A slightly increased amount of epicardial fat was present along the course of the coronary arteries. The heart and great vessels presented no other external abnormalities. The right atrial wall was paper thin and translucent with visible, delicate, widely separated, flattened muscle bundles but with some areas totally devoid of muscle. In the fixed state the consistency of the atrial wall resembled that of parchment. The dilatation involved the free wall of the atrium only and was directed anteriorly, superiorly, and laterally (that is, to the

Figure 3

(A) Postmortem double contrast roentgenogram. Outlined by contrast medium are the anterior leaflet of the tricuspid valve, right ventricle, outflow tract, and pulmonary valve cusps. (B) Explanatory diagram of A.
right while in situ), almost completely effacing the atrial appendage particularly when viewed externally. Internally the atrial appendage was represented by a ring of hypertrophied muscle fibers surrounding a paper-thin circular area, which, however, showed no aneurysmal bulge. A plastic bag introduced into the lumen of the intact right atrium could be filled with 1.5 L of water without overdistending the chamber. The atrial wall adjacent to the ostia of the venae cavae was of normal thickness. Dissection revealed an intact interatrial septum, with a sealed foramen ovale and no evidence of dilatation or bulging of the fossa ovalis. The ostium of the coronary sinus was of normal size. The floor of the right atrium was abnormally flattened, particularly anterior to the tricuspid annulus, forming a shelf representing in part the inner reflection of the deep atrioventricular groove noted externally, in part a fold of redundant atrial wall, and in part the normal floor of the right atrium. The tricuspid ring was normally placed and of normal size but was compressed anteroposteriorly to an elongated slit. Gentle traction readily restored a normal configuration to the tricuspid annulus. The tricuspid valve leaflets could readily be approximated, completely closing the tricuspid orifice.

When the right ventricular cavity was opened, it was apparent that the anterior portion of the roof of the right ventricle also formed the shelflike anterior portion of the floor of the right atrium. This common structure had been displaced downward, presumably by the large volume of blood contained within the right atrium. This had led to a decrease in the space between the anterior leaflet of the tricuspid valve and the anterosuperior wall of the right ventricle.

The right ventricular cavity was moderately dilated, the myocardium averaging 0.3 cm in thickness exclusive of trabeculations. The papillary muscle of the conus received chordae from part of the anterior leaflet, the remainder of the chordae from this leaflet being inserted into three anterior papillary muscles, one small and two larger (fig. 4). The septal and posterior leaflets of the tricuspid valve were continuous and normal. The septal band of the crista was normal, whereas the moderator band was unusually prominent and the trabeculae carneae were moderately hypertrophied. The outflow tract was dilated, with a normally formed crista supraventricularis. The pulmonic valve and pulmonary artery were of normal size and configuration. The left atrium and mitral valve were entirely normal in size and

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**Figure 5**

(A) Postmortem double contrast roentgenogram. Outlined by contrast medium are the left atrium, left ventricle, and aorta. Note how the anterior shelf formed by the right atrium compromises the right ventricle. (B) Explanatory diagram of A.

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appearance. The left ventricle was moderately dilated, its myocardium averaging 1 cm in thickness. The anterior papillary muscle was composed of three partially fused elements, and the posterior muscle consisted of two portions, fused only at the base. Chordae from each mitral leaflet were normally inserted into these. The aortic outflow tract, ascending aorta, and brachiocephalic vessels arose normally (fig. 5). The coronary ostia arose from the aorta. The coronary vessels were free of abnormality.

Microsections of the right atrium revealed that the total thickness of the free wall ranged from 400 microns to 180 microns in diameter and were of normal appearance. Where the atrial wall was at its thinnest, the muscle fibers were widely separated with only a single layer of muscle fibers interposed between endocardium and epicardium. There was no evidence of fibrosis, fatty infiltration, or inflammation. The ventricular myocardium presented no abnormality except for mild muscular hypertrophy.

Discussion

Idiopathic right atrial enlargement is a rare condition, with only 11 other cases reported in the literature. Other authors have emphasized the importance of recognizing the condition as a clinical entity, primarily because these patients may be subjected to exploratory thoracotomy for an erroneous diagnosis of mediastinal tumor. Pastor and Forte, as well as Sumner and associates, stressed the benign nature of the lesion, and indeed all their patients were asymptomatic. However, the patient whose case was reported by Saigusa's group experienced palpitation and fatigue for 21 years and developed atrial fibrillation following exploratory thoracotomy. Following excision of a thin-walled massively dilated right atrium (described as the size of an infant's head) 3 years later, the patient became asymptomatic. Our patient spontaneously developed atrial flutter with fatigue and died suddenly 6 months later. The right atrium also was massively enlarged and predominantly thin-walled. Thus, both patients who became symptomatic had a massively enlarged right atrium and developed atrial arrhythmias.

The postmortem findings confirmed the clinical diagnosis of isolated atrial enlargement. In addition they revealed the early stages of right ventricular compression by the huge right atrium. Both the unusual flattening of the right atrial floor as well as its downward displacement anteriorly are readily explainable as a result of the weight of the large volume of intra-atrial blood resting on the floor of the right atrium. The downward slope of the anterior atrial floor causing compression of the underlying right ventricle simultaneously could have resulted in anteroposterior flattening of the tricuspid annulus from posteriorly directed pressure. If this was indeed the mechanism for these two unusual features then, had the patient lived, the process could have led to progressive right ventricular compression and embarrassment of right ventricular filling as well as possible inflow obstruction at the tricuspid ring.

The postmortem findings of moderate right ventricular compression may help to explain the clinical finding of pulsus paradoxus. Deep inspiration by increasing right atrial blood volume might have caused further compression of the right ventricle with a decrease in right ventricular filling and consequently a further drop in an already abnormally low cardiac output. In a case recently published by Morrow and Behrendt aneurysmal dilatation of the right atrial appendage caused compression of the right ventricle, with a significant decrease in cardiac output shown at cardiac catheterization. This case differs from idiopathic right atrial enlargement only in that the massive enlargement was restricted to the right atrial appendage, whereas in all the other cases, the whole of the right atrium was involved. Indeed, Morrow and Behrendt's case is so similar to those of idiopathic right atrial enlargement reviewed here as to suggest that it represented yet another variant in the spectrum of this entity. Their patient was first noted to have an abnormal cardiac silhouette at the age of 9 years. At age 16, symptoms of palpitations, fatigue, and angina developed. These became progressively worse and were accompanied by syncope. Surgical excision of the atrial appendage 7 years later resulted in complete relief of symptoms. At operation the...
atrial appendage was described as a thin-walled, almost transparent sac, a description that would apply equally well to the whole right atrium, in Saigusa's patient as in our own. In addition the right ventricle was described as "strikingly deformed, having been compressed in its anteroposterior dimension." The four cases reported by Sheldon and co-workers include one diagnosed as right atrial diverticulum. From the description it is not quite clear whether the right atrial enlargement in this patient was due to aneurysmal dilatation or to a pedunculated type of diverticulum. It is noteworthy that like Morrow and Behrendt's patient, this patient was symptomatic and was cured by excision of the right atrial diverticulum. Sheldon and co-workers also remarked on distortion of cardiac chambers by the diverticulum and there apparently was impedance to ventricular filling.

If one were to include Morrow's and Sheldon's cases in the spectrum of idiopathic right atrial enlargement, then three of the 14 patients developed atrial arrhythmias, and one also had a decrease in exercise tolerance and syncopal attacks. A fourth experienced palpitations and signs of progressive right-sided inflow obstruction. Three patients became asymptomatic after surgical excision, while our patient, who refused operation, died suddenly 6 months after the onset of symptoms. All four symptomatic patients had massive enlargement of all or part of the atrium, and as Morrow and Behrendt pointed out, the excessive size of the right atrium could predispose to circus movements and the occurrence of irritable ectopic foci. Indeed, two otherwise asymptomatic patients developed transient atrial fibrillation during cardiac catheterization. Patients with massive atrial enlargement, therefore, should be followed at regular intervals, and early excision of the dilated portion of the atrium seems to be the treatment of choice. Should atrial arrhythmias or syncope occur, surgery should be performed without delay. The possibility that patients with massive enlargement of the atrium may develop compression of the right ventricle before developing arrhythmias or other symptoms is an added reason for considering early surgical excision. The patients reported by Pastor and Forte and Sumner and associates all exhibited only moderate atrial enlargement, and all were asymptomatic. These observations led these authors to conclude that the lesion is a benign one. However, the possibility that atrial arrhythmias or right ventricular compression may develop at a later date even in patients with only moderate atrial enlargement cannot be ruled out. The reported follow-up period in the cases reported by Pastor and Forte and Sumner's group was too short to rule out this possibility.

Because of the pitfalls in diagnosis, previous reports on idiopathic right atrial enlargement have stressed the importance of recognizing this atrial enlargement as a clinical entity. Some of these patients have undergone exploratory thoracotomy because of an erroneous diagnosis of mediastinal tumor. It should be realized that these patients may also present with features suggesting pericardial effusion or tamponade. The enlarged atrium interposed between the heart and anterior chest wall may result in distant heart sounds and a low voltage ECG, as occurred in Morrow and Behrendt's patient, one of Sheldon and associates', and our own. Pulsus paradoxus was present in our patient and transient pericardial friction was heard in two patients. These signs, together with massive enlargement of the cardiac silhouette, may well suggest pericardial effusion.

Of the 14 cases of idiopathic atrial enlargement reported, 10 were in young males and were detected during routine x-ray examination, six of them while serving in the Armed Forces. Two of the four female patients were 9 and 19 years old when cardiomegaly was first discovered by routine x-ray examinations. Both sought help only after they had become symptomatic, 7 and 21 years later. A third female patient, whose condition was detected at the age of 45, refused all further investigation as she was asymptomatic. The fourth female patient was the only one of the entire
group that presented initially with symptoms. Had it not been for routine x-ray examinations while serving in the Armed Forces, the condition would not have been detected in six asymptomatic males and one asymptomatic female. The apparent predominance of males therefore may be a fortuitous one. Considering the circumstances that led to the detection of this abnormality in almost three quarters of the reported cases, one would expect a much higher incidence of the disease than is suggested by the paucity of present reports.

The etiology of this entity has not been clarified to date, and it is tempting to speculate that the continual pressure of a large volume of blood within an already structurally abnormal atrium could lead to progressive dilatation. However, the fact that in our patient little change in size of the atrium occurred over a period of 9 years and that the enlargement was present at the age of 7 years, suggests that the enlargement is not rapidly progressive and that it is most probably a congenital lesion. Other authors \(^1\)-\(^5\) also have noted evidence of atrial enlargement at an early age with no significant change over several years, observations ranging from 2 to 24 years. They, too, have postulated a congenital basis for the lesion.

The paper-thin appearance and quality of the right atrial wall with some areas almost totally devoid of musculature as noted at postmortem examination bears a striking resemblance to the description of cases of so-called "parchment heart" or Uhl's anomaly. In Uhl's anomaly there is marked myocardial aplasia or hypoplasia, or both, involving one or more cardiac chambers. The myocardium may be replaced by fibrous or adipose tissue, or there may be direct continuity between endocardium and epicardium. The involved chamber or chambers are commonly described as being large, parchment-thin, and cystlike.

In Osler's original case of "parchment heart" mentioned in his textbook in 1905, and subsequently described by Segal in 1952, \(^8\) all cardiac chambers showed marked myocardial hypoplasia. In 1952, Uhl \(^9\) described a case of almost total absence of the myocardium of the right ventricle. Since then this anomaly has been termed "Uhl's anomaly" and has been regarded as affecting primarily the right ventricle. Yet on reviewing the eight published cases \(^8\)-\(^14\) it is evident that in only two was the right ventricle solely affected. \(^8\), \(^12\) In three cases both the right atrium and the right ventricle were involved. \(^10\), \(^14\) In one case only the left atrium was spared, \(^11\) and in Osler's original case all chambers showed marked myocardial hypoplasia. \(^8\) The question therefore arises whether right ventricular involvement is a sine qua non of Uhl's anomaly, or whether any chamber may show isolated myocardial hypoplasia. There seems no logical reason to postulate that right ventricular involvement must always occur in conjunction with other chamber involvement, and it is, tempting, therefore, to call idiopathic right atrial enlargement a form of Uhl's anomaly, or parchment right atrium. Certainly both the gross appearance and the microscopic findings are similar to those described in Uhl's anomaly. Aneurysmal dilatation of the right atrial appendage alone as described by Morrow and Behrendt \(^7\) or of part of the right atrium described as diverticulum by Sheldon and associates \(^4\) may represent a form of focal or partial parchment right atrium. Focal myocardial hypoplasia has previously been observed both in the right atrium \(^14\) and right ventricle. \(^12\) Finally, the possibility that some cases of "idiopathic aneurysmal dilatation" of the left atrium or left atrial appendage may also fall into the same category merits consideration. In some an abnormally thin wall with myocardial hypoplasia or replacement by fibrous tissue has been remarked upon. \(^15\)-\(^17\)

In conclusion patients presenting with isolated enlargement of the right atrium, particularly those with massive enlargement, should seriously be considered for prophylactic resection of the atrium to prevent the development of arrhythmias and ventricular compression. Cases of congenital enlargement of the right atrium or atrial appendage may represent a form of Uhl's anomaly, and the
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possibility of involvement of other cardiac chambers should be considered.

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