Idiopathic Enlargement of the Right Atrium

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In 1961 Pastor and Forte reported three patients with enlarged right atria but without significant enlargement of other cardiac chambers or evidence of heart disease. They ascribed a congenital origin to this malformation and named it idiopathic enlargement of the right atrium. Although a previous report alluded to right atrial enlargement in association with right ventricular enlargement, Pastor and Forte’s paper constitutes the first recognition of isolated idiopathic right atrial enlargement. In keeping with the usual trend following the description of newly recognized diseases, it was anticipated that additional cases of idiopathic enlargement of the right atrium would soon be recognized and reported. No confirmatory reports of such an entity have appeared in the English literature, however, and only one in the Japanese literature. Thus, further documentation of this diagnosis appears essential for its recognition and acceptance as a clinical entity. The purpose of the present report is to add four additional cases of idiopathic enlargement of the right atrium, two of which were subjected to detailed investigation in an attempt to define the etiology of this malformation and to establish its existence as a nosologic entity.

Materials and Methods

Cardiac catheterization was performed according to standard technics. Hemodynamic data were recorded with Statham strain gages (P 23 D) coupled to a multi-channel photographic recorder. Blood oxygen saturations were determined with use of standard cuvettes and Van Slyke manometric technics. Indicator-dilution curves were recorded with indocyanine green and a Gilford densitometer. Nitrous oxide tests were obtained according to the procedure of Sanders and Morrow. Carbon dioxide angiography was performed according to the technic of Burch and Phillips. Intracavitary electrocardiography was carried out and interpreted as described by Hernandez et al.

Patients Studied

Patient 1

M.J.J., a 19-year-old Caucasian was referred to the U.S. Naval Hospital for evaluation because of an abnormal chest roentgenogram. He was asymptomatic, and his exercise tolerance was good. There was no past history of rheumatic fever, chest trauma, pericarditis, or significant pulmonary infection. The family history was positive for arteriosclerotic heart disease but no members had congenital heart disease.

The blood pressure was 134/74 mm. Hg and the pulse was 76 per minute. The heart was not enlarged. A systolic ejection sound was audible at the left sternal border. There were no murmurs, and the remainder of the examination was unremarkable.

The routine laboratory studies were normal. The electrocardiogram showed that the terminal QRS forces were directed superiorly and anteriorly, producing a small R prime in V5. The chest roentgenograms demonstrated a prominent right heart border (figs. 1A and 2A).

Because of the unusual cardiac contour, right heart catheterization including exploration of the right heart with an electrode catheter and cineangiography was performed. The right heart pressures, oxygen data, nitrous oxide test, indicator-dilution curves, and an intracardiac electrocardiogram were normal (table 1). Isolated enlargement of the right atrium was visualized during cineangiography (fig. 3). The tri-
cuspid valve appeared to be in a normal position. The final conclusion was idiopathic right atrial enlargement without associated intracardiac defects.

**Patient 2**

J.S.N., a 37-year-old Caucasian had a history of ulcerative colitis of 9 years' duration. Routine chest roentgenograms 2 years previously showed prominence of the right heart border. The patient had had no symptoms suggesting congenital or acquired heart disease. During a recent hospitalization for ulcerative colitis, the prominence of the right atrial border was again noted.

The general physical examination was within normal limits. The blood pressure was 138/82 mm. Hg, and the pulse was regular at a rate of 70. No cardiac thrills or abnormal impulses

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**Figure 1**

Posteroanterior chest roentgenograms. Note the prominent right heart borders. A, patient 1; B, patient 2; C, patient 3; and D, patient 4.
RIGHT ATRIAL ENLARGEMENT

were palpable. The first sound at the tricuspid area was split during inspiration and in the same area a soft systolic ejection sound was noted during expiration. The second sound at the base was physiologically split. Along the left sternal border there was a soft grade I/VI systolic ejection murmur, heard loudest with the patient supine.

The electrocardiogram showed wide S waves in leads II, III, V₅, and V₆. The tracing was considered to be within normal limits. The chest roentgenograms are shown in figures 1B and 2B. Cardiac catheterization was undertaken because of the prominent right heart border. Pressure data obtained in the right heart were within normal limits (table 1). Blood oxygen satura-

Figure 2

Left anterior oblique chest roentgenograms. The prominent right atria are emphasized in this projection. A, patient 1; B, patient 2; C, patient 3; D, patient 4.
tions, indocyanine-green dye curves, and nitrous oxide tests indicated that an intracardiac shunt was not present. Intracavitary electrocardiographic recordings during pullback of the catheter from the right ventricle to the right atrium showed normal changes. A carbon dioxide angiocardiogram was obtained with the patient in the left decubitus position. Figure 4 shows the control and 9-second films from the series displaying a large gas bubble of carbon dioxide in the enlarged right atrium with normal thickness of the right atrial wall and pericardium. Conventional angiocardiography with contrast medium was also performed and confirmed the presence of an enlarged right atrium with a normally placed tricuspid valve.

**Patient 3**

V.D.Y., a 22-year-old Negro was referred to the Cardiology Clinic because of a soft cardiac murmur and an unusual heart shadow on chest roentgenograms. The patient had no symptoms referable to the cardiovascular system. There was no past history of pericarditis, rheumatic fever, scarlet fever, or diphtheria. There were no family members with cardiovascular disease.

The physical examination revealed no abnormalities. The blood pressure was 118/62 mm. Hg. The second heart sound was physiologically split and there was a soft grade I/VI systolic ejection murmur along the left sternal border.

Routine laboratory studies were normal, including a negative sickle-cell preparation. The electrocardiogram was within normal limits with a mean QRS axis of +100°. The chest roentgenograms demonstrated a peculiar right cardiac border (figs. 1C and 2C). In view of the experience with the first two patients, this patient was considered to have idiopathic right atrial enlargement and further studies were not considered indicated.

**Patient 4**

C.W., a 21-year-old Caucasian was referred to the Cardiology Clinic for evaluation of an abnormal chest roentgenogram. The patient denied symptoms referable to the cardiovascular system. Previous physical examinations had been within normal limits, and the patient had not previously received reports of abnormalities of routine chest roentgenograms.

The blood pressure was 116/64 mm. Hg. The entire physical examination was unremarkable. There were no abnormal cardiac pulsations or murmurs. The second sound in the pulmonic area was normally split.

The electrocardiogram was within normal limits. The unusual cardiac silhouette is shown in figs. 1D and 2D. In view of the completely normal electrocardiogram, auscultatory findings, and the radiologic similarity to the previous patients, the enlargement of the right atrium was considered to be unassociated with any intracardiac defect and to represent idiopathic enlargement of the right atrium.

**Table 1**

<table>
<thead>
<tr>
<th>Site</th>
<th>M.J.J. (Patient 1)</th>
<th>J.S.N. (Patient 2)</th>
</tr>
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<tbody>
<tr>
<td></td>
<td>Pressure, mm. Hg</td>
<td>O2 saturation (%)</td>
</tr>
<tr>
<td>SVC</td>
<td>—</td>
<td>69</td>
</tr>
<tr>
<td>RA</td>
<td>(2)</td>
<td>71</td>
</tr>
<tr>
<td>RV</td>
<td>18/4</td>
<td>72</td>
</tr>
<tr>
<td>PA</td>
<td>17/7 (11)</td>
<td>72</td>
</tr>
<tr>
<td>Arterial</td>
<td>136/70 (90)</td>
<td>92</td>
</tr>
<tr>
<td>Nitrous oxide test</td>
<td>3.4% (no shunt)</td>
<td>3.2% (no shunt)</td>
</tr>
</tbody>
</table>

**Figure 3**

Single frame from posteroanterior cinefluorogram of patient 1. The contrast medium was injected into the right atrium via a cardiac catheter. The right atrial cavity is considered to be enlarged. SVC, superior vena cava; RA, right atrium; RV, right ventricle; and PA, pulmonary artery.
Discussion

The diagnosis of idiopathic right atrial enlargement is based on two essential criteria. The first is the demonstration that the right atrium is disproportionately large compared to the other cardiac chambers. A reasonably accurate assessment of this change may be achieved through routine four-way roentgenograms of the chest and cardiac fluoroscopy. This enlargement is even more convincingly demonstrated, however, by positive or negative contrast angiocardiography (figs. 3 and 4). The second criterion and the one necessary before the term “idiopathic” can be applied is the systematic exclusion of all cardiovascular lesions known to produce right atrial enlargement. These include tricuspid valve disease (stenosis or regurgitation), obstructions to right atrial emptying (e.g., tumor), various causes of pulmonary hypertension, pulmonary stenosis (valvular and infundibular), left-to-right shunts (atrial septal defect with and without anomalous pulmonary venous drainage, complete anomalous venous drainage, and incomplete anomalous venous drainage without atrial septal defect), and Ebstein’s anomaly. All of these lesions, especially those of sufficient degree to cause detectable right atrial enlargement, can usually be excluded on clinical grounds alone and detailed laboratory study is not necessary. Two of these lesions, however, may not be so readily diagnosed, viz., incomplete anomalous pulmonary venous drainage without associated atrial septal defect and Ebstein’s anomaly. In the former defect, the anomalous veins may not be easily visualized roentgenographically, the second sound at the pulmonic area may vary normally with respiration, murmurs may be soft or absent, and the electrocardiogram may be normal. Thus, clinical suspicion of the abnormality is difficult and laminography, cardiac catheterization, and angiocardiography may be necessary. Ebstein’s anomaly in its fully developed state is easily diagnosed on clinical grounds but forms frustes of the anomaly associated with right atrial enlargement may be extremely difficult to exclude. In this regard the value of intracardiac electrocardiography in establishing the diagnosis has been emphasized.

An essential criterion for the acceptance of idiopathic enlargement of the right atrium as a nosologic entity is the recognition of a
certain degree of homogeneity in the clinical and laboratory findings of the patients reported. Pastor and Forte's patients, because of associated diseases, may not have been sufficiently convincing examples of the syndrome for widespread acceptance. None of their three patients underwent cardiac catheterization and thus hemodynamic data were lacking. Further, none of their patients was investigated by means of intracardiac electrocardiography. One patient was diagnosed at exploratory thoracotomy for an erroneously suspected neoplasm involving the heart. The right atrium and right atrial appendage were markedly enlarged and dilated. Microscopic examination of the atrial appendage revealed hypertrophy of muscle fibers. In addition a pulmonary nodule was resected and found to be a granulomatous lesion compatible with tuberculosis. A second patient was diagnosed by means of angiocardiography. This patient had multiple congenital skeletal anomalies and bronchoscopic examination disclosed a congenital anomaly of the right bronchial tree. Further, this patient had had a diagnosis of acute rheumatic fever with carditis established 7 years prior to final study. The potential etiologic significance of rheumatic tricuspid valvular deformity in producing the right atrial enlargement was not explored. A third patient refused further study and was diagnosed as idiopathic right atrial enlargement by means of routine chest roentgenograms. In this patient the right atrial deformity had been previously attributed to pleuropericardial adhesions that could not be excluded as the cause of the abnormal cardiac silhouette. Despite the presence of such associated diseases in Pastor and Forte's patients they are probably valid examples of idiopathic enlargement of the right atrium.

More recently a single case report by Saigusa et al. of idiopathic enlargement of the right atrium has appeared in the Japanese literature. The patient, a 43-year-old woman who complained of fatigue and palpitations, was referred with a diagnosis of cardiac aneurysm. A previous thoracotomy had shown a massively dilated heart, and postoperatively there had been transient atrial fibrillation. Physical examination revealed only an increased area of cardiac dullness. The electrocardiogram was normal except for prominent P waves, especially in leads V1 and V6. Cardiac catheterization showed normal right heart pressures and angiocardiography revealed a massively enlarged right atrium. The diagnosis of idiopathic enlargement of the right atrium was confirmed at thoracotomy and the larger part of the dilated area was excised and removed during total cardiopulmonary bypass. Although intracardiac electrocardiography had not been employed, it was considered that the surgical findings excluded anomalies other than right atrial dilation. Microscopic examination of the right atrial wall revealed irregular thickness and distribution of cardiac muscle fibers.

The four patients reported herein, because of the lack of associated diseases and uniformity of clinical findings, confirm and strengthen the notion that this disease represents a distinct clinical entity. None of the patients had a past history suggestive of cardiopulmonary disease nor did any have symptoms attributable to the enlarged right atrium. In all, the auscultatory findings were normal as were the electrocardiograms. The absence of symptoms, physical findings, and electrocardiographic abnormalities in our patients confirms the absence of such findings in the patients of Pastor and Forte. Whether any of these seven patients will progress to develop symptoms or electrocardiographic abnormalities similar to the patient of Saigusa et al. remains to be seen. The similarity of chest roentgenograms in the seven patients is also remarkable. All were referred for cardiac evaluation because of an abnormal right heart border observed roentgenographically. Right atrial enlargement was confirmed by contrast angiocardiography in two of the patients. In addition two of the patients were subjected to cardiac catheterization and intracardiac electrocardiography. Pressure determinations, oxygen data, nitrous oxide tests, and dye-dilution curves were within normal limits, excluding significant intracardiac
shunts, valvular regurgitation, or obstructive lesions. Intracardiac electrocardiography excluded the possibility of Ebstein's anomaly.

From the uniformity of clinical and laboratory findings cited above, it is believed that the diagnosis of idiopathic right atrial enlargement can be made on clinical grounds alone. Further, it is concluded that this disease represents a specific clinical entity. The cause of such an abnormality, however, is completely in doubt. Evidence is not available to support either a congenital or acquired origin, but the young age and absence of history of cardiovascular disease in our patients supports the former. It is also possible that a common etiology does not exist in all patients. Hemodynamic studies have shown no deviation from normal and no intracardiac pressure gradients have been detected. Nor is there evidence to support the existence of an abnormal sequence of activation or contraction of the involved chamber. Finally the term "enlargement" when applied to the cardiac chambers may refer either to increase in muscle mass of the wall (hypertrophy), to increase in chamber volume (dilatation), or to both. From the angiographic studies, the predominant structural change implied is that of dilatation. Right atrial biopsy of the patients subjected to thoracotomy showed hypertrophy of muscle fibers in one case and irregular thickness and distribution of cardiac muscle fibers in the other. Whether this is a primary feature or secondary to initial dilatation remains to be determined. Idiopathic dilatation of the right atrium has not yet been recognized at postmortem examination.

It is important to recognize idiopathic right atrial enlargement as a clinical entity. In some such patients, right atrial enlargement may be correctly diagnosed but erroneously attributed to significant intracardiac defects. In others, the abnormal right heart border may be considered to result from pleural or pericardial adhesions, defects, cysts, or, more importantly, neoplasms adjacent to or involving the heart. That two of the four previously reported patients have undergone diagnostic thoracotomy lends emphasis to the need for further recognition and acceptance of idiopathic enlargement of the right atrium as a distinct nosologic entity.

Summary

Four patients with idiopathic enlargement of the right atrium are described. All patients were evaluated carefully by clinical means and two were subjected to extensive investigation including cardiac catheterization, angiocardiography, and intracavitary electrocardiography.

The diagnosis of idiopathic enlargement of the right atrium is based primarily on radiographic examination with exclusion of other causes of right atrial enlargement by appropriate clinical and laboratory means. In the patients studied no cause for the right atrial abnormality could be established. Nonetheless, it is concluded that this disease is a distinct clinical entity that deserves further recognition and investigation.

References

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