The Significance of the Left Atrial Appendage in Rheumatic Heart Disease

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SUMMARY The possible diagnostic value of an enlarged left atrial appendage (LAA) on the posterior-anterior or right anterior oblique chest film as a means of implicating a rheumatic etiology for mitral valve disease in children was investigated. Chest films were examined without prior knowledge of clinical or laboratory data, and the results were later correlated with this information in 113 children and adolescents. The clinical and laboratory data included application of the modified Jones criteria for the diagnosis of acute rheumatic fever, streptococcal antibody titers and clinical and cardiac catheterization findings.

In children with mitral valve disease, our data suggest that an enlarged LAA, especially in the presence of pulmonary venous obstruction, is characteristic of rheumatic heart disease. This finding appears to be particularly useful, in conjunction with streptococcal antibody studies, in distinguishing rheumatic from nonrheumatic patients with mitral insufficiency.

OVER THE PAST SEVERAL YEARS, it has been noted that a dilated left atrial appendage on the posterior-anterior (PA) chest film was frequently associated with rheumatic endocarditis of the mitral valve.1-2 This observation was based on experience with numerous cases of rheumatic and nonrheumatic disease of the mitral valve in both children and adults.3 Although the body of the left atrium often showed enlargement in patients with nonrheumatic mitral valve disease, the left atrial appendage (LAA) was rarely enlarged. This finding raised the possibility of using an enlarged LAA on radiologic examination as an adjunct in the diagnosis of rheumatic heart disease. Supporting data to validate this impression were therefore sought.

Recently, the use of certain streptococcal antibody tests [anti-streptolysin O (ASO), anti-desoxyribonuclease B (anti-DNAse B), and anti-group A carbohydrate (A-antibody)] in distinguishing rheumatic from nonrheumatic mitral valve disease in children was refined.45 The presence of an elevated A-antibody with normal ASO and anti-DNAse B was recently reported in patients with rheumatic valvular disease but not in congenital mitral insufficiency.6 The importance of correctly identifying those patients with mitral valve disease of rheumatic origin relates to the success of anti-streptococcal antibiotic prophylaxis in the prevention of recurrent rheumatic attacks. However, there is little advantage and significant risk in treating patients with mitral valve disease of nonrheumatic etiology in the same fashion. The risk includes both the potential hazard of anaphylactic reactions and the possible higher incidence of bacterial endocarditis with penicillin-resistant organisms in patients receiving oral penicillin.

With the availability of these streptococcal antibody tests for separating rheumatic from nonrheumatic mitral valve disease, this joint study was undertaken to determine the value of a dilated LAA in diagnosing the etiology of mitral valve disease. This communication reports the results of analysis of the radiologic finding of a dilated LAA as a means of implicating a rheumatic etiology for mitral valve disease in a group of 113 children and adolescents.

Material and Methods

Patients

The case material included 113 patients evaluated in the Rheumatic Fever Clinic at the University of Florida College of Medicine. Age, sex, and follow-up data are summarized in table 1. All patients had ASO, anti-DNAse B, and A-antibody tests performed on their initial visit and at frequent intervals during follow-up. On the basis of clinical and laboratory information, this initial patient population was divided into four groups: Group 1 — children with mitral valve disease during and/or following a well-documented attack of acute rheumatic fever; Group 2 — children with mitral valve disease without history of acute rheumatic fever (ARF) but with patterns of streptococcal antibody titers characteristic of chronic, inactive rheumatic mitral valve disease;5 Group 3 — children with congenital mitral valve disease or mitral valve disease of nonrheumatic etiology; and Group 4 — children with documented clinical and laboratory evidence of ARF without evidence of heart disease.

Group 1 included 43 patients who manifested mitral valve disease at the time of initial presentation, 42 with mitral valve insufficiency and one with mitral valve stenosis. Seven patients had murmurs of aortic valvular insufficiency as well. All 43 patients presented with an acute illness which fulfilled the modified Jones criteria5 for the diagnosis of ARF. Each patient in Group 1 had elevated levels of streptococcal antibodies. Follow-up in 35 patients showed a fall of
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in congenital aortic valve lesions of ASO and anti-DNAse B titers toward normal with persistence of elevated A-antibody titers, the pattern previously found to be associated with chronic rheumatic valvular disease.

Group 2 consisted of seven patients without historical evidence of ARF who were referred for evaluation of possible rheumatic heart disease because of auscultatory findings of mitral insufficiency or stenosis. Two cases had aortic insufficiency as well. All seven showed elevated A-antibody levels in association with normal ASO and anti-DNAse B titers and were presumed to have rheumatic heart disease.

Group 3 consisted of 50 patients with nonrheumatic mitral valve disease. None of these patients manifested an acute illness fulfilling the modified Jones criteria. Thirty-eight patients had negative streptococcal antibody titers while 12 showed evidence of recent streptococcal infection, i.e., elevated ASO, anti-DNAse B titers and A-antibody levels. The titers in each of these 12 patients showed a nonrheumatic response, returning to normal within a year. Forty-nine of these patients had mitral insufficiency while one had mitral stenosis. The 50 patients in this group included 15 with endocardial cushion defects and mitral insufficiency, ten with mitral insufficiency associated with other congenital heart lesions. These included patent ductus arteriosus, atrial septal defect, ventricular septal defect, and congenital aortic valve lesions in varying combinations. Ten additional patients were asymptomatic, presented with mitral insufficiency without history of ARF, and had normal streptococcal antibody titers. Nine patients had mitral insufficiency shown to be associated with a prolapsing mitral leaflet, in six patients by right and left heart catheterization and angiocardiography and by echocardiographic documentation in the remaining three. Five patients had mitral insufficiency associated with myocardiopathy while one patient had congenital mitral stenosis associated with aortic stenosis and coarctation of the aorta. Thirty-nine of the 50 patients were studied by cardiac catheterization and angiology.

Group 4 consisted of 13 patients who fulfilled the modified Jones criteria for acute rheumatic fever but who had no evidence of cardiac involvement on initial presentation or during follow-up. All 13 patients had elevated streptococcal antibody titers with subsequent fall to normal.

Radiologic Analysis

In order to determine the significance of LAA enlargement in rheumatic heart disease, plain films were analyzed in the following manner. Chest films and/or cardiac series of the 113 patients of Groups 1-4 were randomized and evaluated independently by two cardiac radiologists without knowledge of clinical and laboratory information. A cardiac series included four views of the heart with barium in the esophagus. Analysis included assessment of the following parameters:

A. Pulmonary Vascularity. The pulmonary vascularity was determined to be normal or increased. Increased vascularity was characterized as either pulmonary venous obstruction (pulmonary venous hypertension or congestion) or shunt vascularity. Pulmonary venous obstruction (PVO) and shunt vascularity were further graded as mild, moderate, or severe depending on previously described and well-established criteria.

B. Heart Size. The heart size was graded using a 0 to 4+ classification. Because of the variabilities inherent in a system employing exact measurements of heart size, the

<table>
<thead>
<tr>
<th>Patients investigated</th>
<th>No</th>
<th>Male</th>
<th>Female</th>
<th>Age in years at presentation</th>
<th>Years of follow-up</th>
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</thead>
<tbody>
<tr>
<td><strong>Group 1</strong> — Rheumatic heart disease</td>
<td>43</td>
<td>23</td>
<td>20</td>
<td>Median (range)</td>
<td>Median (range)</td>
</tr>
<tr>
<td><strong>Group 2</strong> — Presumptive rheumatic heart disease</td>
<td>7</td>
<td>3</td>
<td>4</td>
<td>9.4 (6-18)</td>
<td>3.85 (1-7)</td>
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<tr>
<td><strong>Group 3</strong> — Nonrheumatic mitral valve disease</td>
<td>50</td>
<td>17</td>
<td>33</td>
<td>7.3 (0.5-18)</td>
<td>4.3 (1-10)</td>
</tr>
<tr>
<td><strong>Group 4</strong> — Acute rheumatic fever without heart disease</td>
<td>13</td>
<td>5</td>
<td>8</td>
<td>11.0 (6-17)</td>
<td>2.4 (1-3)</td>
</tr>
</tbody>
</table>

Figure 1. Lateral views of the chest with barium in the esophagus showing degrees of enlargement of the left atrial body. A) Normal left atrial body. B) 1+ left atrial enlargement. C) 2+ left atrial enlargement. D) 3+ left atrial enlargement.
observers evaluated the heart in four views and graded size on the basis of their own experience. Mild cardiomegaly was represented by 1+ and 4+ represented severe, gross enlargement of the cardio-pericardial silhouette.

C. Size of Left Atrial Body. The criteria for determining enlargement of the left atrial body was based on the degree of indentation of the barium-filled esophagus in the PA, RAO, and lateral views. In addition, elevation of the left main stem bronchus was used in the PA and LAO views. The 0 to 4+ classification reflected primarily the barium-filled esophagus in the lateral and RAO views. This was graded as follows:

0 — no interruption in the contour of the esophagus at the level of the left atrium (fig. 1A).
1+ — barely perceptible posterior displacement and anterior interruption of the esophageal contour at the level of the left atrium in an esophagus that is not overly distended with barium (fig. 1B).
2+ — posterior displacement with interruption of the anterior and posterior esophageal contour, easily appreciated as rather abrupt changes from segments above and below the left atrium (fig. 1C).
3+ — marked posterior displacement of the esophagus at the level of the left atrium with distinct changes from the course of the esophageal segments above and below the left atrium (fig. 1D).
4+ — this includes the so-called giant left atrium syndrome. There may be esophageal escape with the barium-filled esophagus deviated to the left and a large mass protruding to the right of the spine. In addition, there may be compression of the left main stem bronchus resulting in impaired ventilation of the left lower lobe.

D. Left Atrial Appendage. The size of the LAA was graded on a 0 to 3+ classification. Left atrial appendage enlargement was determined to be present on the basis of the appearance of a convex bulge in the characteristic position of the left atrial appendage in the PA and/or RAO views. This was graded as follows:

0 — normal left heart border (fig. 2A).
1+ — a barely perceptible convex bulge in the region of the LAA (fig. 2B).
2+ — a distinct convex bulge in the region of the LAA (fig. 2C).
3+ — an overt, often eccentric bulge in the region of the LAA (fig. 2D).

More than one series of films were reviewed in 46 of the 50 patients in Groups 1 and 2 and in all 63 patients in Groups 3 and 4. After analysis of the patients' films, decisions were made as to the presence or absence of an enlarged LAA. Only then were clinical and laboratory information correlated with the X-ray findings.

Results

Groups 1 and 2 (50 patients)

These two groups of patients with rheumatic or presumptive rheumatic heart disease are combined for ease of presentation.
Incidene and Degree of Left Atrial Appendage Enlargement on Any Given Examination. Of the 50 patients with rheumatic or presumptive rheumatic heart disease, 38 (76%) had an enlarged LAA (table 2). Of these, 27 showed 1+ enlargement, ten showed 2+ enlargement, and one showed 3+ enlargement (fig. 2D). The 12 lacking enlargement of the LAA will be dealt with separately (see false negatives).

Changing Size of Left Atrial Appendage on Follow-up Examinations. Twelve of the 38 patients with an enlarged LAA demonstrated a change in size on follow-up examination, with nine decreasing in size, and three becoming larger. Among the 27 patients with 1+ LAA enlargement, seven lost evidence of LAA enlargement over a period ranging from 1 to 7 years (fig. 3), and in five of these seven patients, the mitral insufficiency murmur diminished in intensity or completely disappeared (2 cases). In all seven patients streptococcal antibody titers returned to normal levels in the same period. Six of these seven patients demonstrated concomitant diminution in size of the body of the left atrium. Follow-up studies in two of the ten patients with 2+ LAA findings revealed loss of evidence of LAA enlargement. In both instances, mitral insufficiency murmurs decreased in intensity and the size of the left atrial body decreased.

Three patients who initially had LAA enlargement showed an increase in LAA size during follow-up. In one patient the small change noted was judged to be due to variation in size seen at the time of atrial systole and diastole (fig. 4). In two other patients, progressive increase in LAA size was seen over 5 years and 9 months, respectively (fig. 5). In each instance the mitral insufficiency murmur increased in intensity, the body of the left atrium enlarged, ASO and anti-DNAse titers diminished and A-antibody level remained elevated.

Relationship of the Left Atrial Appendage to Pulmonary Vascularity. Of the 50 patients with rheumatic or presumptive rheumatic heart disease, 41 showed evidence of some degree of PVO on at least one occasion (table 3). In the

**Table 2. Presence of Enlargement of Left Atrial Appendage in Study Patients**

<table>
<thead>
<tr>
<th>Patients investigated</th>
<th>Enlarged LAA</th>
<th>Negative LAA</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Group 1 — RHD</td>
<td>31</td>
<td>12</td>
<td>43</td>
</tr>
<tr>
<td>Group 2 — Presumptive RHD</td>
<td>7</td>
<td>0</td>
<td>7</td>
</tr>
<tr>
<td>Group 3 — Nonrheumatic MV disease</td>
<td>2</td>
<td>48</td>
<td>50</td>
</tr>
<tr>
<td>Group 4 — ARF without heart disease</td>
<td>1 (?)</td>
<td>12</td>
<td>13</td>
</tr>
</tbody>
</table>

**Figure 4.** Posterior-anterior chest films in one of three patients who appeared to demonstrate an increase in size of the LAA. This patient had rheumatic mitral insufficiency. A) A barely perceptible convex bulge (white arrows) is seen in the region of the LAA. B) The convex bulge in the region of the LAA (white arrows) is slightly more prominent on a film taken a short time later. This case is thought to demonstrate change in LAA size with atrial diastole.

**Figure 5.** Posterior-anterior chest film with barium in the esophagus demonstrating an increase in size of the LAA over a five year period in a patient with progressive rheumatic mitral insufficiency. A and B) PA chest films at age 9 and 14 years both demonstrate severe pulmonary venous obstruction. The LAA was initially graded at 1+ and subsequently at 3+ (white arrows). C and D) Lateral films with barium in the esophagus show progressive enlargement of the left atrial body from 1+ to 3+ (black arrows).
remaining nine, pulmonary vascularity was interpreted as normal on all examinations. LAA enlargement was seen in 36 of the 41 patients with PVO (88%) (table 4). More significant, however, is the fact that 36 of the 38 (95%) patients with enlarged LAA had associated mild to severe PVO (table 4). In the nine patients with normal vascularity, only two had an enlarged LAA. In both cases the LAA grading was initially 1+ and returned to normal on subsequent follow-up exams. Two of the seven patients without evidence of LAA enlargement or PVO manifested enlargement of the body of the left atrium. Six of the seven had clinical evidence of very mild mitral insufficiency while one patient had predominant aortic insufficiency with minimal mitral insufficiency (fig. 6).

Patients with Rheumatic Heart Disease Lacking Enlargement of LAA: False Negatives. Of the 12 patients with rheumatic heart disease without an enlarged LAA, seven showed normal vascularity and heart size. Six of these seven had very mild or transient mitral insufficiency while the seventh had predominant aortic insufficiency with minimal mitral insufficiency. Clinically, all seven of these patients were determined to have rheumatic heart disease of mild degree. Five rheumatic patients had negative LAA despite the presence of PVO. One had fulminant rheumatic carditis, massive pericardial effusion with a globular-shaped heart (fig. 7A), and died three days after hospitalization. Another patient had severe chronic mitral insufficiency and aortic insufficiency with probable pericardial effusion (fig. 7B), eventually requiring surgical replacement of the mitral and aortic valves; it is interesting to note that radiologic evidence of the LAA appeared transiently during the postoperative period. In both of these cases with pancarditis the LAA was hidden by the pericardial effusion. The ability of a pericardial effusion to mask a dilated LAA is shown in figure 8. The three additional patients had predominant aortic valve disease in the presence of mild mitral insufficiency (fig. 9), which suggests that when rheumatic endocarditis affects primarily the aortic valve, little LAA dilatation may result.

Size of the Left Atrial Body and Correlation with Left Atrial Appendage Enlargement. Forty-three of the 50 patients with rheumatic or presumptive rheumatic heart disease had radiographic enlargement of the body of the left atrium. Thirty-seven were in Group 1 and six were in Group 2. This enlargement was graded as 1+ in 31 patients, 2+ in ten patients, and 3+ in two patients. In general, there was good correlation between the size of the left atrial body and the size of the LAA. The exceptions included three of the 38 patients with LAA enlargement who manifested normal left atrial body size, in each case associated with mild PVO. Three other patients had 1+ enlargement of the left atrial body without enlargement of the LAA. The only other instances of lack of correlation included the two cases of severe carditis with pericardial effusion discussed above.

Group 3 (50 patients)

Incidence of Left Atrial Appendage Enlargement. All 50 patients in this group had some form of nonrheumatic mitral valvular disease. Left atrial appendage enlargement was seen in only two of these patients. One was a 2-year-old female with associated ventricular septal defect, atrial septal defect, and 4+/4+ mitral insufficiency. Although her appendage was 1+ in size at 2 years and 4 years of age, it was normal by age 5 (fig. 10). The other patient was an 18-month-old female with a partial form of endocardial cushion defect and
4+/4+ mitral insufficiency. Her LAA was 2+ in size (fig. 11). Fortunately, these patients could be readily distinguished from patients with rheumatic mitral valve disease by the presence of shunt vascularity on plain film examination rather than evidence of PVO. Enlargement of the LAA in these two patients was likely related to the nonspecific hemodynamic effects of their severe mitral insufficiency and the consequent gross left atrial enlargement.

Size of Left Atrial Body. Thirty-five of the 50 patients with nonrheumatic mitral disease had radiographic enlargement of the body of the left atrium. This enlargement was graded as 1+ in 23 patients, 2+ in eight patients, and 3+ in four patients. This breakdown of left atrial body size is similar to the patients in Groups 1 and 2 with rheumatic or presumptive rheumatic heart disease. Both patients with nonrheumatic disease who manifested an enlarged LAA also had 3+ enlargement of the left atrial body.

Group 4 (13 patients)

Incidence of Left Atrial Appendage Enlargement. Among the 13 patients with clinical and laboratory evidence of ARF without evidence of endocarditis, 12 had no evidence of LAA enlargement while one showed questionable enlargement of the LAA on two occasions four years apart. All other radiologic and clinical aspects of the cardiac exam were normal in this patient (fig. 12). No patients showed enlargement of the left atrium, cardiomegaly or abnormal pulmonary vascularity.

Discussion

It has been emphasized in recent studies that a significant proportion of mitral valve disease, especially in children, may be of nonrheumatic etiology. Differentiation of rheumatic from nonrheumatic disease in the young patient presenting with mitral valve disease without clear-cut history of ARF may be difficult. The usefulness of certain streptococcal antibody tests in aiding in this distinction has been suggested in a recent study. The present work was undertaken to determine whether a finding on plain chest film, namely enlargement of the left atrial appendage (LAA), might also be of use in this regard.

With few exceptions both the radiologic literature and textbook descriptions of the radiographic findings in rheumatic heart disease have failed to stress the possible diagnostic value of the dilated LAA. Some investigators have discussed the enlarged LAA in connection with the so-called mitral configuration, and have included LAA enlargement as a criterion for generalized left atrial enlargement. Kaye et al. reported the presence of a dilated LAA in 41 of 100 adults and children with mitral valve stenosis alone and in combination with other valve lesions. He found it to be present in three cases of mitral stenosis without other radiological evidence of left atrial enlargement. The etiologic documentation in these patients is uncertain but assumed to be rheumatic. Jacobson and Weidner stated that the dilated LAA was one of the most reliable manifestations of mitral valve disease, and suggested performance of the Valsalva maneuver as a means of making the LAA more visible. They theorized that the rheumatic process made the appendage more distensible than normal and, therefore, visible following release of the Valsalva maneuver. Interestingly, they encountered seven patients in whom demonstration of a temporarily dilated LAA was the only positive radiographic finding of mitral valve disease.

Two uncommon entities characterized by radiographic

![Figure 8](http://circ.ahajournals.org/)

**Figure 8.** Posterior-anterior chest films showing the ability of a pericardial effusion to mask a dilated LAA. A) PA chest film on a 12-year-old girl with acute rheumatic pancarditis and mitral insufficiency. There is a moderate degree of pulmonary venous obstruction and an enlarged globular-shaped heart. B) Follow-up film three months later shows little improvement in the pulmonary venous obstructive vascularity, but a definite LAA is now seen (arrows).

![Figure 9](http://circ.ahajournals.org/)

**Figure 9.** Posterior-anterior chest roentgenograms in two of 5 patients with significant rheumatic heart disease and absence of an enlarged LAA. Both patients had 3+/4+ aortic valve insufficiency and minimal mitral valve insufficiency. A) Twelve-year-old girl with mild pulmonary venous obstruction, a prominent pulmonary trunk but no definite LAA. B) Thirteen-year-old boy with mild PVO, an enlarged left ventricular contour to the heart and no definite LAA enlargement.
evidence of isolated LAA enlargement were recognized prior to the onset of this study. These include congenital pericardial defects which may be total or partial\textsuperscript{18-27} (total being the more common) and idiopathic LAA dilatation with intact pericardium, a rare condition associated with supraventricular tachycardia and systemic emboli.\textsuperscript{28-30} Other common to rare conditions which may simulate the radiologic appearance of a dilated LAA include (a) congenital diverticulum of the left atrium,\textsuperscript{31} (b) juxtaposition of the atrial appendages, (c) aneurysm of the right ventricular infundibulum, seen primarily in postoperative tetralogies, (d) cardiac tumors in the area of the LAA, (e) aneurysm of the ascending aorta or pulmonary trunk, (f) single ventricle with ventricular inversion and (g) mediastinal tumors. It cannot be overemphasized that the vast majority of the above conditions will present radiologically with a pulmonary vasculature picture other than PVO.

Our past experience has emphasized the fact that enlargement of the LAA has been rare to uncommon in the wide range of nonrheumatic cardiac lesions which can lead to enlargement of the body of the left atrium. The 50 patients in Group 3 with nonrheumatic mitral valve disease include a wide variety of lesions which may lead to left atrial enlargement. These include left-to-right shunts with intact atrial septum, endocardial cushion defects, congenital mitral insufficiency, nonobstructive myocardial infarction, prolapsing mitral valve and asymmetric septal hypertrophy. Only two of these 50 patients (4\%) showed evidence of LAA enlargement, both cases associated with left-to-right shunts (figs. 9 and 10). These cases emphasize the importance of considering the nature of the radiologic changes in pulmonary vascularity in interpreting the presence of an enlarged LAA.

Kaye reported a dilated LAA to be present in only one of 62 patients without heart disease\textsuperscript{4} while Jacobson and Weidner found only one false positive LAA in several hundred patients who had congenital and acquired nonrheumatic heart disease.\textsuperscript{5}

Overall, the data presented indicate that there is a very good correlation between the presence of a dilated LAA and clinical and laboratory evidence of rheumatic mitral valve disease. This association is particularly striking when radiologic assessment of the pulmonary vascularity is considered. Left atrial appendage enlargement was found in 36 of 41 (88\%) patients with rheumatic mitral disease severe enough to cause some degree of PVO. It was primarily in those patients with very mild or transient rheumatic mitral valve disease that the LAA was not frequently seen. In addition, there was excellent correlation between serial changes in the size of the LAA and the severity of mitral insufficiency as judged by clinical criteria and levels of streptococcal antibody titers. Among the seven patients (Group 2) presenting with mitral insufficiency without history of ARF but with a pattern of streptococcal antibodies similar to those previously described as characteristic of patients with chronic inactive rheumatic heart disease,\textsuperscript{4,5} each manifested LAA enlargement. This finding further supports this radiologic criterion as an aid to the differentiation of rheumatic and nonrheumatic mitral disease.

The patients in Group 4 who had clinical and laboratory evidence of rheumatic fever without rheumatic endocarditis represent a group which essentially acts as normal controls. It is intriguing that one of these patients showed

\[\text{Figure 10. Posterior-anterior chest roentgenograms in one of two patients with nonrheumatic mitral valve disease who demonstrated LAA enlargement. This patient has a ventricular septal defect, an atrial septal defect, a right arch with an aberrant left subclavian artery, and 4+4+ mitral valve insufficiency. A) Film taken at age 2 demonstrating mild shunt vascularity with a 1+ enlarged LAA (arrows). B) Film at age 4 demonstrating continued prominence of the vascularity and an increase in the heart size but no definite LAA. Note the right aortic arch (arrows). In patients with shunt vascularity, the LAA has no significance since children with rheumatic heart disease show either normal vascularity or PVO.}\]

\[\text{Figure 11. Posterior-anterior chest film and anterior-posterior left ventriculogram in an 18-month-old female with an endocardial cushion defect. A) Chest film at age 18 months showing moderate prominence of the vascularity of the shunt type (not well reproduced here) and 2+ LAA enlargement (arrows). B) Left ventriculogram demonstrating 4+/4+ mitral valve insufficiency with opacification of a markedly enlarged left atrium (LA) and a dilated left atrial appendage (LAA).}\]
questionable enlargement of the LAA (fig. 12). Could the rheumatic process have involved the appendage in this patient without involving the mitral valve?

Inasmuch as our study deals with growing children, the effects of thoracic growth on the relative size of the cardiac chambers should be mentioned. The ratio of cardiac chamber size to the thoracic volume is increased in infancy and childhood and this ratio gradually diminishes as individuals grow older (as the thoracic cage increases in size). Therefore, it might be assumed that chest films taken in infancy and childhood may be more sensitive in detecting volume variations in various chambers of the heart.

The mechanism of LAA enlargement in rheumatic heart disease, particularly when the mitral valve is involved, is unlikely to relate merely to hemodynamic factors and changing heart-thoracic volume ratios. This is apparent from the low incidence of dilated LAA in nonrheumatic conditions which result in enlargement of the body of the left atrium. Rather, it appears likely that the LAA is dilated in rheumatic heart disease as a consequence of myocardial inflammation, particularly in the region of the LAA. That the LAA myocardium may be particularly susceptible to such inflammation could relate to the fact that the LAA, like the mitral valve, is embryologically derived from cardiac tissue while the body of the left atrium is derived from the extracardiac common pulmonary vein.

The data presented here suggest that the finding of radiologic evidence of an enlarged LAA in a child with mitral valve disease, especially in association with evidence of PVO, is a reliable sign indicating a rheumatic etiology for the mitral valve disease.

References

2. Elliott LP: A Roentgenologic Approach to Heart Disease. New York, Medcom, 1974

Table 4. Correlation between Pulmonary Venous Obstruction Pattern and Evidence of LAA Enlargement among Study Groups 1 and 2

<table>
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<tr>
<th></th>
<th>PVO</th>
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<tr>
<td>LAA +</td>
<td>36</td>
<td>2</td>
</tr>
<tr>
<td>LAA -</td>
<td>5</td>
<td>7</td>
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![Figure 12. Posterior-anterior chest film of a patient with clinical and laboratory evidence of rheumatic fever but without evidence of rheumatic endocarditis. This patient demonstrated a questionably enlarged LAA (arrows) on two chest films taken 4 years apart.](image)
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