Case Report

CONGENITAL LEFT ATRIAL ENLARGEMENT
A CASE REPORT WITH SPECIAL REFERENCE TO
MYOCARDIAL FINE STRUCTURE

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Intrapericardial congenital left atrial enlargement is regarded as a rare condition. Since Palacio's description in 1960,1 about 10 cases have been reported in the English literature.1−9,33 The possibility that the intrapericardial congenital left atrial enlargement may also fall into the same category of idiopathic right atrial enlargement or Uhl's anomaly, was considered.10 But, the etiology of these anomalies is not clear. Some histologic examinations of the affected atrium were described but myocardial fine structure of the congenital left atrial enlargement has not yet been reported.

The purpose of this report is to describe a patient with intrapericardial congenital left atrial enlargement who also has a moderate enlargement of the right atrium. This represents the first case in which the electron microscopic findings of the myocardium are available.

Case Report

The patient was a 3-year-old girl who was suffering from measles. On a routine chest roentgenogram, the presence of abnormal cardiac silhouette was observed. She has been healthy and asymptomatic without any abnormal past history such as rheumatic fever or myocarditis.

Key Words:
Congenital left atrial enlargement
Left atrium
Electron microscopy

(Received on June 25, 1974; Accepted for publication on October 28, 1974)
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The right atrium was also moderately enlarged. The left ventriculogram showed no evidence of mitral regurgitation.

Open heart surgery was performed. The pericardial sac was intact. The left atrium was aneurysmally enlarged and the right atrium also showed moderate dilatation. No clot was found inside the dilated atrium. The foramen ovale was patent but no valvular incompetence of the foramen ovale was noticed on open heart observations. A small left-to-right shunt at the atrial level was thought to pass via the insufficiency of the patent foramen ovale because of the dilated atrium. A part of the aneurysmally dilated left atrium was excised. The post-operative course was uneventful.

**Materials and Methods**

The specimens of the left atrium, right atrium and right ventricle were obtained during the open heart surgery. Each specimen was light and electron microscopically examined. Conventional staining with hematoxylin eosin and monoamine fluorescence technique of Falck and Hillarp were
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Fig. 3. Left atriograms show markedly dilated left atrium.

Fig. 4. Longitudinally sectioned muscle fiber of the left atrium shows disarrangement of myofibrils and an abnormal Z band (arrow). X 7500

carried out. For electron microscopy, the samples were fixed in cold 5% glutaraldehyde in phosphate buffer for 2 hours, washed in the same buffer for 24 hours, refixed in 1 per cent buffered osmium tetroxide for a hour and embedded in epoxy resin according to the method of Luft. Thin sections were cut with a Porter Wulm MK-1 ultramicrotome, stained with uranyl acetate and lead nitrate, and then examined with a Hitachi HU-11A electron microscope.

Observations
On light microscopy, the left atrial wall was
thin but showed a normal cardiac structure without any signs of inflammation or fibrosis. The histochemical demonstration of sympathetic innervation of both atria showed decreased monoaminergic nerve fibers among myocardiocytes especially on the left atrium.

On electron microscopy, important changes affecting subcellular organelles were seen in the myocardium of the left atrium especially on mitochondria and myofibrils. Mitochondria were markedly increased in number. The large part of the left atrial cardiocyte was occupied by numerous clusters of mitochondria. The increase of mitochondria was especially prominent at the perinuclear region and under the sarcolemma but, they were also increased among myofibrils apart from nucleus. (Fig. 4, Fig. 5) However, the size and the morphology of these mitochondria were normal, and no swelling of the matrix or shrinkage of the cristae were observed. Mitochondrial granules were sometimes observed.

Myofibrils were decreased. Disorganization of the uniform arrangement of myofibrils parallel with each other, was often seen. In comparison with normal cardiocytes, myofibrils run in a
Fig. 7. Abnormal Z bands.

Fig. 8. Crista-like organelle (arrow). ×24000

Fig. 9. Numerous large atrial granules with some deformity are seen near the Golgi complex. ×12600
disorderly way in the various directions, not always along the long axis of the muscle cells. (Fig. 4) Contraction bands were often observed sometimes in parallel with neighbouring cells. (Fig. 6)

Especially interesting observations were the structural alterations of the myofibrils in the Z bands. Frequently, Z band appeared as electron dense irregular thickening sometimes extended up to the length of one or more sarcomeres. Distortion and dispersion of the widened Z bands were often observed. (Fig. 7a, b, c.) In other areas, cristal like organelles having a fine cross-striation with a regular repeating period of 200 Å were observed between myofibrils near the contraction band. (Fig. 8) The abnormal Z bands resembling these were reported by Bishop and Cole.

Atrial specific granules were increased in number among the cluster of mitochondria in the left atrial cardiocytes. The sizes of the granules were various and some granules were enlarged up to 0.6 µ in diameter. Irregular shaped granules were sometimes observed. (Fig. 9) The sarcoplasmic reticulum was dilated sometimes up to the large vacuoles.

The right atrium also showed same changes to some degree and right ventricular myocardiocytes showed increased mitochondria, but, abnormalities of the myofibrils were not observed.

DISCUSSION

Congenital enlargement of the left atrium was divided into intra- and extrapericardial types. However, these two types are thought to be two separate disease entities representing atrial and pericardial conditions. The extrapericardial type is primarily a deficiency of a portion of the pericardium through which the left atrium may herniate. The intrapericardial type is thought to be caused by congenital abnormalities of the left atrial wall. On electron microscopic examinations of the left atrial cardiocytes in this case, mitochondria were markedly increased in number, but, myofibrils were decreased and large parts of the cardiocytes were occupied by mitochondria. On experimental left ventricular hypertrophy with aortic constriction by Page, the increase in myofibrillar volume occurred and mitochondrial fraction of cell volume decreased. Similar decreases of the ratio of mitochondria to myofibrils were obtained in cardiac hypertrophy. In the experimental congestive heart failure, increase of the myocardial mitochondria in number was observed; however, the size of the mitochondria was decreased and morphological changes of the mitochondria such as swelling of the matrix or shrinkage of the cristae were observed. Pease found the increase of the amount of mitochondria in idiopathic cardiac hypertrophy in man and named it mitochondriosis. These observations were confirmed by other investigators. But, mitochondrial damages also occurred in these conditions.

Palade noted a decrease in the ratio of mitochondrial to myofibrillar mass in the hypertrophied auricular appendages from patients with lesions of the mitral valve. On the contrary, our case showed a marked increase of mitochondria without any morphological changes. These facts show that this left atrial enlargement is neither due to cardiac hypertrophy nor to congestive heart failure and may be due to some abnormalities of the myocardium itself.

According to Meerson's experimental observations of left ventricular myocardium on hyperfunction, morphological changes were described as follows. During the emergency stage of hyperfunction, substantial alterations occurred in mitochondria. The area occupied by swollen mitochondria was increased and the area occupied by myofibrils was correspondingly reduced. During the stage of relatively stable hyperfunction, myofibrils also increased and the area occupied by mitochondria was smaller than in earlier periods. There were few signs of mitochondrial destruction. During the stage of gradual exhaustion, an increase in the rate of destruction and a reduction in the mass of mitochondria were characteristic. The mass of the mitochondria of the myocardial cell indicates the potential efficiency in the production of energy rich substances necessary for contraction. Our case showed abnormal Z bands, decrease or disarrangement of myofibrils, and increase of morphologically normal mitochondria. These findings may suggest that myofibrillar hypofunction due to abnormal myofibrils exists in the congenital left atrial enlargement and the reduced myofibrillar function results in mitochondrial increase as in the stable stage of Meerson's experiments without myofibrillar increase.

The abnormal Z bands observed in this left atrial muscle cells resemble the rod-like structures in skeletal muscles of patients with nemaline myopathy. These abnormal Z bands were recently reported in the experimentally hypertrophied right ventricles of dogs and in

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the right ventricle of an old male cat, and these abnormal Z bands were thought to be identical with tropomyosin. Hodge reported about very thin cristals of tropomyosin, and a striking resemblance between his cristals and cristal-like structures observed in our case also suggests this structure to be tropomyosin. Tropomyosin is thought to be an inhibitor of myofibrilar ATP-ase. Depression on the activity of ATP-ase is correlated with a depression of myocardial contractility. The decrease of contractility due to abnormal myofibrils may have some relation to the dilatation of left atrium.

Other abnormal observations were concerned with the atrial specific granules. In normal human atrium, atrial specific granules are small in size with average diameters of 0.25 μ. In our case, left atrial cardiocytes contained very large granules 0.6 μ in diameter. The number of the granules was increased. These specific granules are thought to be closely related to the function of the atrial autonomic nervous system of the heart, and these changes in our case, with decrease in the catecholaminergic nerve fibers on the affected atrium, shows some abnormalities on the left atrial autonomic nervous functions.

The fact that the right atrium also showed the same changes may suggest that these abnormalities affect not only the left atrium but also both chambers to some degree.

**Summary**

A case of intrapericardial congenital left atrial enlargement was reported. The patient is an asymptomatic 3 year-old girl with abnormal cardiac silhouette seen on a chest roentgenogram. Angiography showed marked left atrial enlargement, and the diagnosis of intrapericardial congenital left atrial enlargement was confirmed on operation. Specimens of the atria were electron microscopically examined. Myocardiocytes of the left atrium showed marked increase of the mitochondria and decrease of myofibrils. Abnormal Z bands and disarrangement of myofibrils were often observed. Atrial specific granules in the left atrium were increased in size and number. The right atrium also showed same changes. The pathogenesis of this condition was discussed.

**References**


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