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EX AEDIBVS ACADEMICIS IN CIVITATE VATICANA

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ENDOCARDIAL FIBROELASTOSIS

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SVMMARIVM — Etsi sunt, qui, cum fibroelastosi endocardica laborent, ad iuvenilem aetatem vel etiam adultam perveniant — at plerique intra primum vitae annum moriuntur — non idem tamen videtur esse ille morbus in pueris atque in adultis, sed duo diversi morbi, ex diversis causis provenientes: id concludunt Auctores, post fibroelastosis endocardicae examen in viro quodam et in quibusdam pueris.

In these last twenty years a particular attention has been devoted to a number of cases of cardiac hypertrophy in which autoptic findings revealed a marked fibrotic thickening of the inner third of the ventricular wall, usually on the left side.

Such observations are not quite recent but only recently the endocardial fibroelastosis in adult subjects has been distinguished from similar conditions formerly confused with it.

LOEFFLER [1] reported on two cases of such a disease using the expression « Fibroplastic chronic endocarditis with eosinophyly ». Some other cases with or without eosinophyly have been reported by PUDDU et al. [2], [3], MUMME [4], SMITH and FURTH [5], ROULET [6], EDGE [7], LENNOX [8], BERBLINGER [9], LENEGRE and GERBAUW [10], MCKUSICK and COCHRAN [11], MCNICOL [12], MARCOLONGO and TUVERI [13],

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LYNCH and WATT [14], VAN BUCHEM, ARENDS and SCHROEDER 15], HOSMAN [16], VAN UEBLINGER et all. [17], GOYER and BOWDEN [18]. Similar conditions have been related by DAVIES and BALL [19] and by SHAPER and WRIGHT [20] in Uganda where the 14 per cent of death occurred during the years 1951-53 and labeled as « cardiac insufficiency » can be ascribed to the above mentioned disease.

MARINI and PINCA [21] have been able to reveal in the majority of cases of fibroelastosis a left ventricular prevalence connected with a clear change of the S-T tract and T wave with a light and changeable reduction of the R wave voltage in left precordial recordings.

DECOURT [22] has reported on interesting data obtained by means of bioptic specimen of myocardium in cases of fibroelastosis; he also related on surgery (flaying of the left ventricular endocardium) presently performed by the cardiosurgical equipe of San Paulo's Medical Clinic.

Up to date the connection passing between adults and children fibroelastosis is not yet cleared up. Some children suffering from the disease can undoubtely reach the teen and even the adult age, though deaths generally occur during the first year of life. However some morphological features could allow a difference between the two conditions.

Having had the opportunity to carry out an anatomic study on a case of fibroelastosis in a male subject of 52 years of age and on two children we will report on the most meaningful characteristics of the cases observed.

FIRST CASE — *G.R., 52 y. o.*

Previously admitted into the hospital owing to a cardiocirculatory insufficiency. Newly admitted (August 1957) because affected by dyspnoea, cyanosis, oedema of lower limbs.

Poor general conditions, semi-orthopnoic posture, cyanosis

of skin and visible mucosae. Symmetric and poorly expanding chest, superficial breath at a rate of 32 per minute. On the basis small and medium buble rattles. Enlargement of the cardiac area with ictus at the fifth space externally to the emiclavear line; right edge two centimeters from the margino-sternal line at the fourth intercostal space. Cardial pulse quite arythmic owing to an atrial fibrillation with a frequency of 96 per minute. Blood pressure, 120/70 mm of Hg. The liver reaches, by means of its rounded lower edge, the transversal umbilical line; it shows an increased consistency and is slightly painful. Conspicuos oedema of lower limbs. Light hypochromic anemia in lack of eosinophyly.

An E.C.G. showed an atrial fibrillation arythmia, with signs of left ventricular prevalence and of coronaric sub-endocardial ischemia.

The patient's conditions remained unchanged till the death occurred on the fourth day; the worsening was associated with a progressive increase of dyspnoea and cyanosis.

The following are the autoptic data concerning with the heart; it presents an increased volume (620 gr) and its distal part is essentially formed by the left ventricle. Once opened the ventricular cavities it has been possible to find out a thickening of the endocardium corresponding to the heart tip, to papillar muscles, to septal wall, to the posterior wall of both the ventricles. Such a thickening has a white mother of pearl colour and in some areas it is extended throughout a third of the whole wall. Hypertrophy and dilation of both the ventricles the walls of which present sclerotic phenomena, peculiar of the sub-endocardial myocardium. The right coronary artery presents an additional opening; the investigation of the sub-epicardial root of coronaric vessels shows atherosclerotic changes represented by fibrotic and ateromatic spots. A careful examination, performed using thin nylon wires, did not reveal occlusions of visible branches.

The histological investigation has been carried out on fragments of both the ventricles. The sections have been stained according the following methods: hematossillin-eosin, VAN GIESON, MALLORY, MALLORY-AZON, WEIGERT.

The histological study, in accordance with macroscopic features, shows in the inner third of the myocardial wall of both the ventricles, a thickening formed by a fibro-connectival dense and homogeneous tissue invaded by focuses of hyalin and metachromatic degeneration. In such areas it is possible to see a number of neoformed vessels forming a vascular gang closed to the sub-endocardial myocardium. In some areas such vessels appear in form of gaps limited by an endotelial layer. Among the connectival tissues some muscle fibrocells appear to be prey of regressive phenomena.

Elastic structures with a rich network of neoformed vessels prevail within the myocardial layer. Such structures appear to be formed by thickened and single fibres or by thin bundles.

The connections between the fibrous layer and sub-endocardial myocardium are changeable. In some areas, the fibrous part appears to be indipendent from the muscolar one, while elsewhere it is possible to see large connectival links, coming from the fibrous layer and piercing, throughout large areas, the myocardium. Such connectival files derange the organization of muscle bundles, taking up a lobar feature. A lot of muscle fibres of the inner layers restrained by the connectival bundles undergo regressive process and then a full destruction. The sub-endocardial vessels show poor changes consisting on periavventitial sclerosis while as to the intramyocardial ones they do not present any significative change.

SENCOND CASE — *C.M., 1 y. o.*

Normal delivery. During the pregnancy the mother suffered from intermittent haesantematic digressions. Normal growth. Whooping-cough at two months of age; boils at five.

Ten days before hospitalization the child suffered from a cold, followed, two days later, by cough, the access of which increased during the night. Poor feverish reaction, marked dyspnoea, noticeable anorexy and depression. The death occurred a few hours after the admission. The autoptic study shown: a number of bronchopneumonic focuses in lower lobes of both the lungs. Heart's volume totally increased. The left ventricle appears markedly expanded with a thickened and whitish endocardium; such a thickening is particularly marked at the upper third of anterior and lateral walls. Dilation of the right ventricle with a whitish endocardium, markedly thickened in the upper third of the anterior and lateral walls. Valvular sets unaltered. No changes of origin and course of coronaric vessels.

THIRD CASE — *R.P., 7 y. o.*

Weight at birth 3000 mr. Good growing up process till the fourth month of age; at this period a progressive decay was setting in; it was associated with a marked hyopchromic anemia. General conditions were successively getting worse. No fever. The patient suffered from a marked anorexy. Dyspnoea and death that occurred a few hours after the addmittance into the hospital during an apshyctic crisis joined with cyanosis.

Autoptic findings: marked cyanosis of lips and finger tips. Moderate asymmetric deformity of the chest owing a precordial pool. Heart totally increased in volume. Once opened the cardial cavities no changes of valvular sets could be revealed. The left ventricular endocardium is markedly thickened so as to assume a tendineous feature; thickening is emphasized at the septal wall and in an area corresponding to the anterior wall. The sub-endocardial layers of myocardium show a more or less advanced sclerosis. Slight hypertrophy of the right ventricle. No septal lacks. The aorta, normal as to the calibre, does not show changes of the intima.

The hystology shows the same findings in all the cases. The abnormal thickening of the endocardium, including in the 2nd case both the ventricles and in the 3rd one only the wall of the left ventricle, results to be formed by a number of elastic fibre bundles with a changeable course, by essentially paralleling the myocardial surface. For a lesser amount, the thickening results from collageneous fibres, set as single ones or collected into small bundles. The reticular stroma is largely represented. In some area, practically in the inner ones, the elastic fibres undergo regressive process consisting on homogeneization and hyalin focuses. The sub-endocardial myocardium often shows a more or less diffused sclerosis with connectival bundles dividing the muscular fibrocells. No changes of sub-endocardial and intramyocardial vessels.

A careful examination of medical litterature does'nt give rise to the possibility of a real settlement of cardial fibroelastosis, namely of the aged form. The clinical picture though not constant and characteristic seems to be that of a cardial progressive insufficiency with a rapid course occurring in young subjects (far below 40 years of age) but sometimes in a more advanced age.

First of all between the childohood and adult age form it is possible to establish some differences starting form a morphological point of view.

In children the endocardial thickening is more extended, particularly in areas corresponding to the sub-aortical part of the ventricular wall; such a thickening is often associated with other deformities (abnormal origin of coronaric vessels, aortic stenosis etc.). The elastic stroma plays an important part in forming the thickening, while as to the collagenous fibres they have not determining roles in such a process.

In adult subjects thickening is more localized in trabecular and tip areas; it is often associated with cavities thrombotic phenomena and consists on a dense fibrous tissue in lack of a plentiful elastic one. However there are some cases in adults

showing the same characteristic of those observed in young subjects.

Higher difficulties can be encountered when tempting to define the aetiopathogenetic moments playing a role in the fibroelastosis determination.

The synthetic aims of our paper do not allow us to report on the different theories adopted to explain the pathogenesis of chilhood fibroelastosis. However, as to this point we have to recall the works of PRIOR and WYATT [23], BLUMBERG and LEYON [24], GOWING [25], ROSSI [26], GROETTINGER, HARVEY, KRAKOVER, OGLESBY [27], STILL [28].

LOEFFLER [1] has included the adult forms in allergic mechanisms and this owing the findings of eosinophyly observed in two cases; the above mentioned A. believes that the anatomoclinical picture is supported by three fundamental features: clinical picture seeming a constrictive pericarditis, eosinophyly, endomyocardial sclerosis, but only a few of the following cases could be settled according these principles. Myocardium and endocardium have been often proposed as initial site of the pathological change. Thickening has been also taken in consideration as end-result of the organization of a wall thrombosis; such hypothesis could be allowed by the feature of the pathological changes that can be regarded as a granulation tissues often associated with an endocavity thrombus.

Furthermore the adult fibroelastosis could be compared to the so-called sub-endocardial infarction during the organization stage; this has emphasized the importance of coronary ischemia in the genesis of fibroelastosis.

The cardiac hypetrophy should represent the initial aetiological phase while the endocardial change would be a consequence of an inadequate blood supply.

The findings of conspicuous endocardial thickening connected with other diseases (beri-beri, pernicious anemia, myocarditis with different aetiologies, lues etc.) have induced LYNCH and WATT [14] to conclude, and this opinion is shared by

MARCOLONGO [13], that the adult fibroelastosis represents the end result of a pathological process having at its begining a lot of causes. The question if the endocardial thickening may be considered responsible of the serious picture of cardial insufficiency observed in such cases has still to be cleared up.

FIGURES

FIGG. 1-2 — First case. Left ventricle showing a marked thickening of the inner layers.

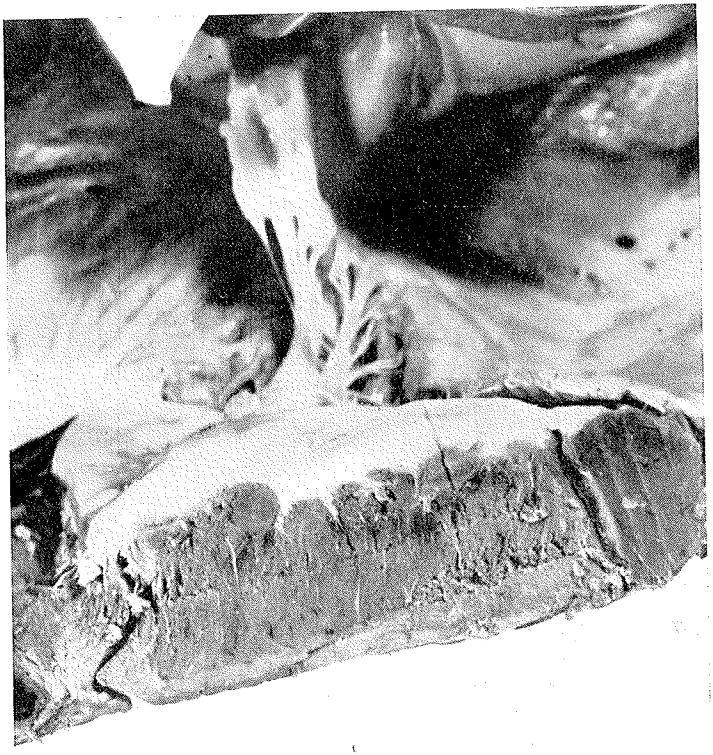

FIG. 3 — First case. Particular view of a left ventricle cut.

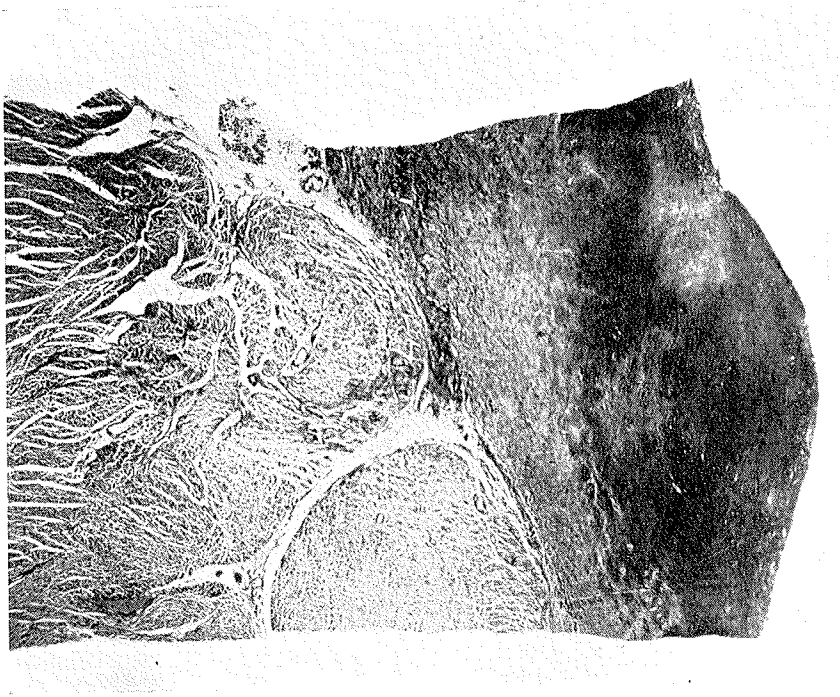

FIG. 4 — First case. Whole microscopic view of the wall of the left ventricle. MALLORY's staining: 10 X.

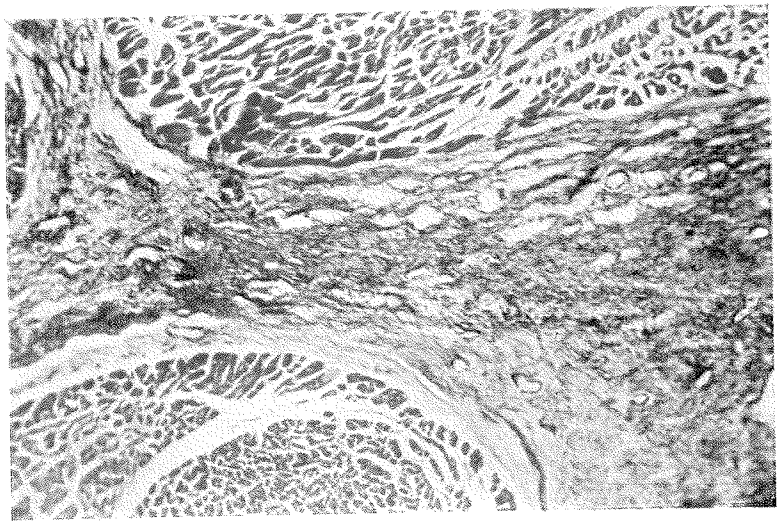

FIG. 5 — First case. The section concerns the passage between myocardium and the endocardial fibrotic area.

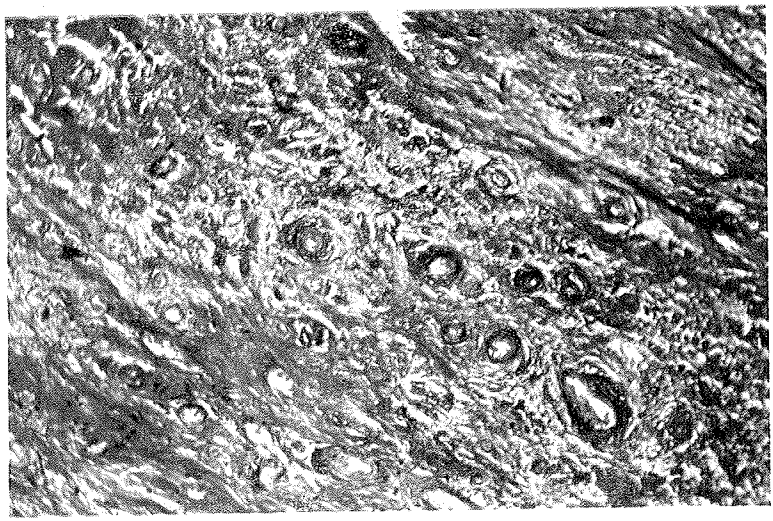

FIG. 6 — First case. The section concerns the passage between myocardium and fibrotic endocardial area; see the number of neoformed vessels. MALLORY-AZAN'S staining; 100 X.

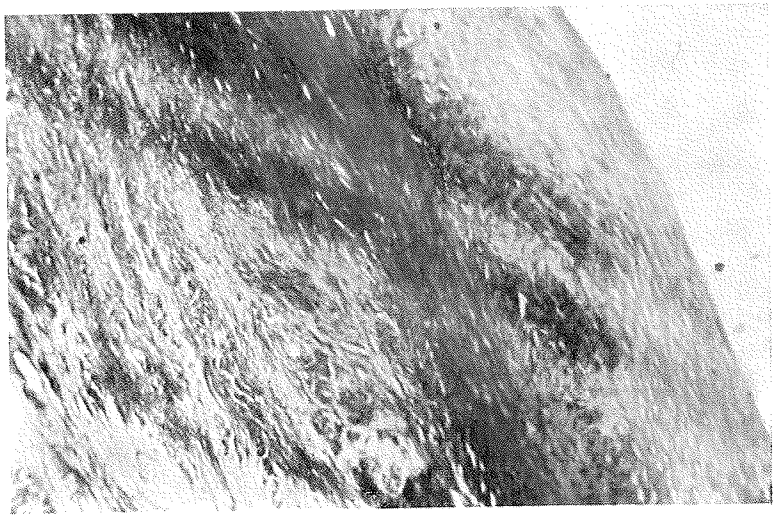

FIG. 7 — First case. Outer layers of the endocardial fibrotic area. MALLORY-AZAN's staining.

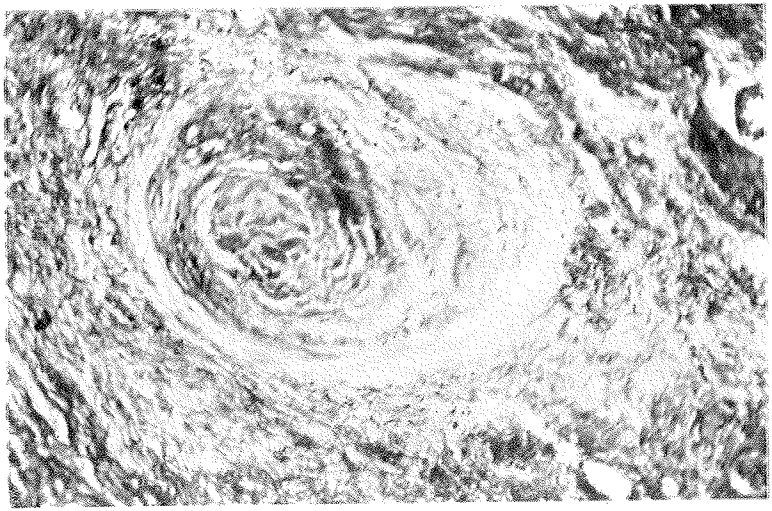

FIG. 8 — First case. Inner layers of the endocardial fibrotic area; see the number of elastic fibres and neoformed vessels. MALLORY-AZAN's staining; 200 X.

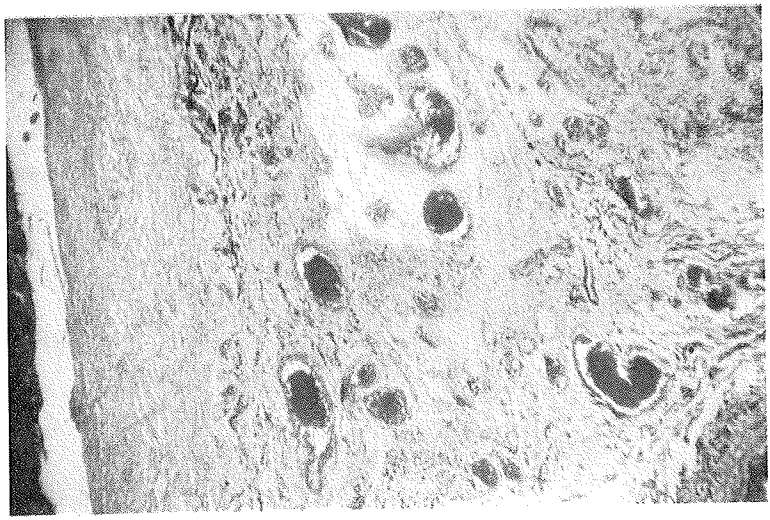

FIG. 9 — First case. Particular view of the previous picture. MALLORY-AZAN staining; 200 X.

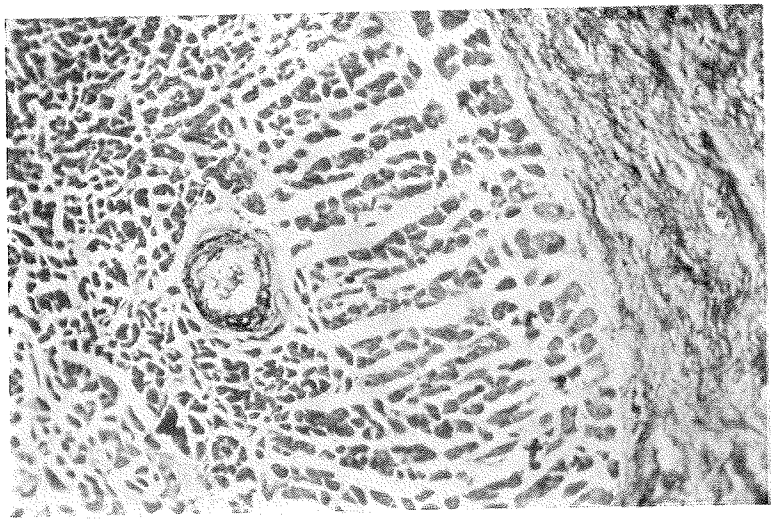

FIG. 10 — First case. Large connectival sept starting from the fibrotic area and dividing the muscle bundles. MALLORY-AZAN staining; 100 X.

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