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# Uhl's Disease: Partial Parchment Right Ventricle

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*Summary.* This is an account of the morpholgical findings of a case of Uhl's disease. In the following is to be found a compilation of all publications which have appeared up to now in world specialist literature and which refer to case aspects of this disease.

Uhl's reflections concerning the morphogenesis of the partial parchment heart are disputed with the aid of some pertinent objections and a new theory is presented which explains the formal genesis.

*Zusammenfassung.* Es wird über die morphologischen Befunde eines Falls von Uhl's Disease berichtet. Im Anschluß daran erfolgt eine Zusammenstellung aller bisher in der Weltliteratur erschienenen, dieses Krankheitsbild betreffenden Publikationen.

Die Uhlschen Überlegungen bezüglich der Morphogenese des "Partial Parchment Heart" werden anhand einiger Einwände widerlegt, und es wird eine eigene, die formale Genese erklärende Theorie vorgelegt.

In 1952 Henry S. M. Uhl published the patho-anatomical findings on the heart of an 8-months old girl who had died with all the clinical signs of a protracted right cardiac failure. In this case the most important feature was the fact that the myocardium was almost completely missing in the area around the right ventricular inlet tract. As in the area in question no morphological signs could be found either of a circulatory disturbance ever having taken place or of a previous inflammation, Uhl drew per exclusionem the conclusion that this abnormality was a matter of congenital deformity.

Despite the fact that observations based on similar evidence had already been recorded (Osler, 1905; Segall, 1950) and that Castleman (1952) almost contemporaneously gave an account of a nearly identical case, this anomaly, to which previously little attention had been paid, has been known from this time forth as Uhl's disease (Uhl's anomaly). The visual analogy gave rise to the frequent use of the rerm "Partial Parchment Heart" for the same disease.

## New Case History

*Hedwig St., married; age: 71.* The cardiac anamnesis of the patient covers a time-span of about 5 months. In May 1973 after having suffered a myocardiac infarct with varying degrees of right and intermittent left bundle branch block (questionable Morgani-Adams-Stokes-attack), she was admitted to the University Medical Clinic Heidelberg (Prof. Dr. G. Schlettler) for pacemaker implantation. The ecg showed signs of an earlier infarct of the anterior wall and a more recent infarct of the posterior wall.

On 25th May 1973 the pacemaker was fitted with myocardial electrodes. Postoperative a visible worsening of the haemodynamic and respiratory condition took place despite normal stimulation of the pacemaker. Peripheral edema and kidney failure developed. The patient died with all the clinical signs of left heart failure.

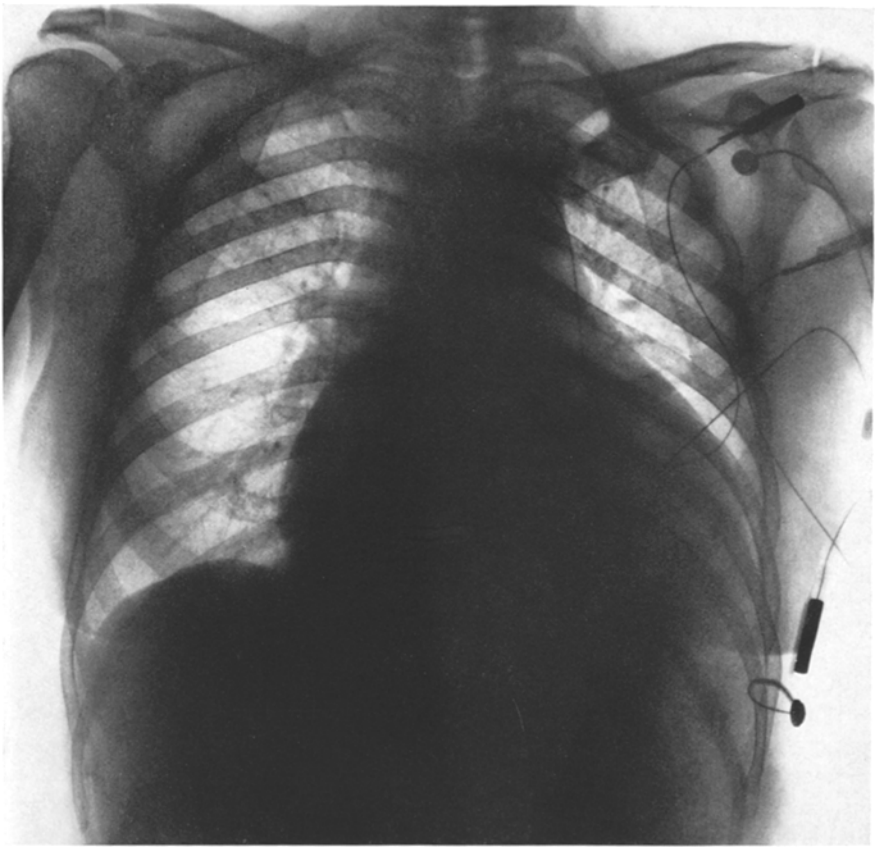

Fig. 1. Chest-X-ray taken shortly before death

## Abstract of the Autopsy Protocol

(Autopsy no. 609/73, Pathological Institute University Heidelberg)

Weight of the heart: 480 grams. The apex of the heart is formed by the right ventricle. The epicardium is superficially covered with fibrinous layers. The places where the myocardiac electrodes were implanted into the heart are not subject to irritation.

Right atrium and ventricle are dilated out of all propertion. The posterior wall of the right ventricle is 2 to 3 mm thick; large sections of the muscular tissue have been replaced by parchment like connective tissue in an area measuring 7 to 5 cm.

The orifices of the cavity veins are as normal. The formaen ovale is closed. The dorsal tricuspid velum is hypoplastic and about 5 mm out of position towards the ventricle. The chordae tendineae are only 1–2 mm long, so that with an ostium measuring 16 cm in circumference a distinct tricuspid insufficiency is present. The septal and anterior vela are normal in form and developement. The muscular conus arteriosus pulmonalis and the pulmonary ostium also give no cause for special attention. The slight hypertrophy of the muscular walls given rise to a deviation of the left cavities towards narrowness. There is also

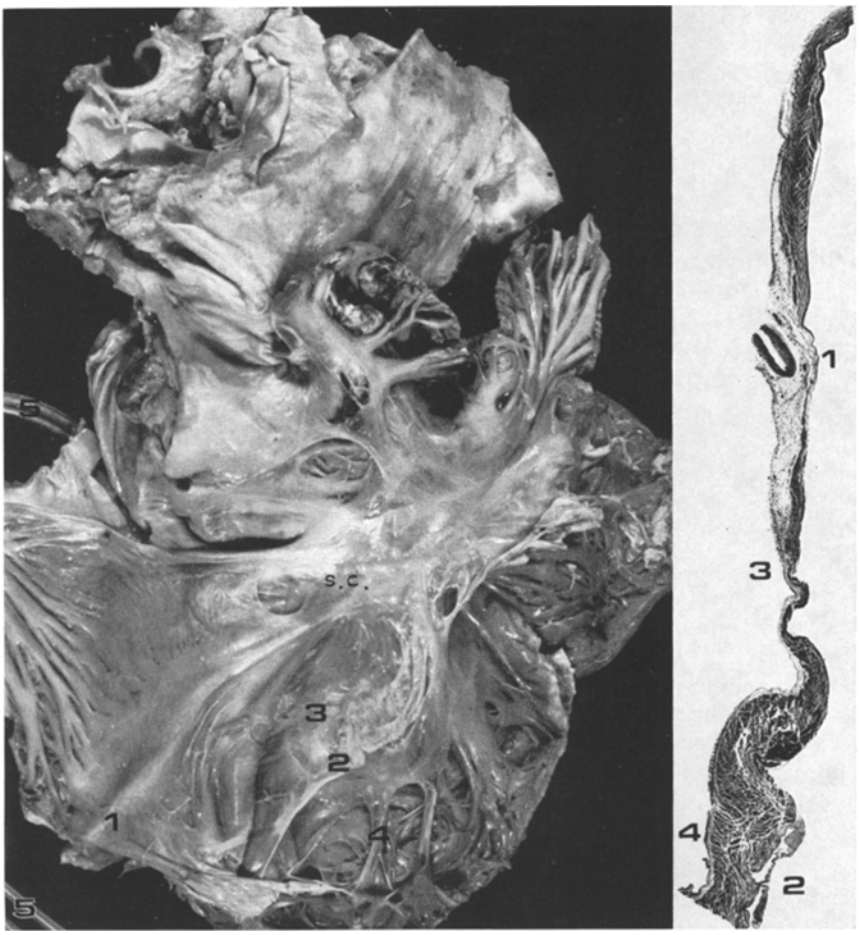


Fig. 2. a Opened right atrium and right ventricle. b Histological specimen of the right posterior atrioventricular region. *s.c.* Sinus coronarius; *1* atrioventricular border (arteria coronaria dextra); *2* towards the ventricle displaced dorsal tricuspid velum; *3* atrialisated region of the posterior free wall of the right ventricle; (Parchment Region); *4* normal ventricular myocardium; *5* pacemaker-electrodes

present an old endocarditis of the mitral valve of moderate degree and a parietal thrombotic formation at the apex of the left ventricle.

The A. coronaria sinistra is hypoplastic to a high degree, whereas the A. coronaria dextra is excessively over-developed and has spread far over onto the posterior wall of the left ventricle. The ramus interventricularis anterior is extremly stenotized. The sectional plane through the apex of the left ventricle shows an old scar as well as several new, clay-colored necroses.

## Histological Evaluation

After the heart had been treated in formalin, samples of tissue were taken from 12 different places for histological examination. Of the seperate preparations

one coloration of haematoxylin-eosin, one of elastica-van Gieson and one of Masson-Goldner were made up.

The most important findings are as follows: In the region of the sinus node is a subtotal fibrosis with cellular displacement action. The circulus sinu-auricularis is hyperaemic and its walls are thickened.

When a section is made through the right atrio-ventricular region the most important and immediate finding is the "parchment region" so typical for Uhl's disease. Here the myocardium consists of few fascilcles of muscular fiber, which are slightly hypertrophied and which lie betwen the epi- and endocardium.

The area around the place where the pacemaker electrodes had been implanted shows extensive fibronecrosis together with central calcination and a serious accompanying myocarditis.

In the hypertrophied left side ventricular wall are scattered perivascular callused inflammations. In the ventricular muscle as well as in the whole area of the atriums signs can be seen of Anitschkow cells.

The infundibula of both coronary arteries show concentric sclerosis and are constricted by cushion-like beds.

As well as the extensive infarct-areas of the left ventricular wall there is an almost diffuse hypoxaemic damage of the inner layer.

## Diagnosis

1. Uhl's disease ("partial parchment heart").
2. Disturbance in the developement of the ostium atrioventriculare dextrum.
3. Right coronary type: a high degree of stenotized calcinatory sclerosis of the ramus interventricularis anterior. Ventroapical and septal myocardiac infarct with old scars on the left ventricular apex.
4. Old, recurrent, rheumatic endomyocarditis with development of numerous, spindle-shaped, perivascular myocardiac scars.
5. Condition after implantation of myocardial pacemaker-electrodes onto the area of connective tissue in the posterior wall of the right ventricle.
6. Acute fibrinous epicarditis.
7. Cardiac pulmonary edema, congestive bronchitis, left pleural effusion.
8. Chronic congestion of the parenchymatous organs lying in front of the right side of the heart.
9. Signs of shock in the kidneys, arterio-arteriolo sclerosis.

## Discussion

An almost complete lack of the myocardium of a ventricle without patho-anatomical signs of any cause whatever is generally accepted to be a relatively rare observation. Since Uhl's publication in 1952 only 31 further cases have been reported in the whole of world specialist literature [1, 3, 5, 6, 8, 17–19, 26, 29, 31–40, 43, 44, 49, 53]: (for compilation see Table 1). Admittedly we must in the context with Uhl's disease certainly reckon with a large number of cases which have never publicly come to light, since in by far the greatest number of cases the correct diagnosis is only made by the pathologist in a post-mortem. As the criteria for a differential diagnosis of this cardiac disease vary very greatly and are relatively uncertain, it is easy to understand how incorrect diagnosis can be made such as Fiedler's myocarditis, Ebstein's anomaly, transposition of the

great arteries, right cardiac failure, glycogen store disease, fibroelastosis or the Bland-White-Garland syndrome.

The over-all picture is made even more complicated by the fact that Uhl's disease can in no way be construed in its aspects as unified. The life-expection of the patient varies considerably from case to case. The most obvious interpretation of this phenomenon would be that the gravity of the clinical consequences is basically dependent upon the size and the localisation of the parchment region in the right ventricle—in other words: upon the degree of the haemodynamic effects.

While Segall (1950) in his pathogenetic reflections concerning the "parchment heart" sees the probability of a myocardiac dystrophy in the context of a generalized muscular dystrophy, and in addition puts forward "elements of a cor pulmonale" to discussion, and Froment (1968) on the other hand tends to classify Uhl's disease between "familiar cardiac muscle diseases" and cases of myocardiopathy, which show "extensive foci of sclerosis without coronary stenosis", Uhl himself (1952) believes that we have here a deal with a congenital defect of the myocardiac anlage of the prospective right ventricle. He supports his arguments concerning the morphogenesis of this anomaly basically on the findings of Davis (1927) and places the time of the teratogenetic determination in circa the 4th somite-stage of the human embryo. According to his hypothesis a pathogenic agent would meet the right-hand section of the paired primitive epimyocardiac mantle before it fuses with the contralateral system and would induce in this way the myocardiac deformity which is strictly limited to the final right ventricle.

This form of the morphogenetic interpretation of the partial parchment heart, which reminds somewhat of the thesis inaugurated by Bredt in 1936, dealing with the "antimeral atrophy" is generally accepted by the authors listed below who have published works on Uhl's disease: Novak (1957), Reeve (1964), Neimann (1965), Gould (1967), Bansi (1968), Sugiura (1970).

With reference to the conceptions of the morphogenesis of a normal and a deformed heart which are valid at the time of writing, however, Uhl's theory must be opposed for the following reasons:

1. A primarily paired system of the primitive myoepicardiac mantle does not exist in mammals (Kl. Goerttler, 1958). In contrast to amphibians and birds we find in mammals only a uniform mesoderm plate above the anterior intestine which, however, divides 2 seperate, secondarily fused, endocardiac tubes from the ventral.

2. Because of a general lack of knowledge concerning the exact fate of the primitive antimera of the human cardiac system (Doerr, 1955) it seems to be impossible to assign the areas of the primitive myoepicardiac mantle to definitively located structures of the differentiated parts of the heart. Moreover, the fact that fundamentally in each ventricle parts of proampulla, of metampulla, of the auditory canal region and of the bulbus tube are to be found together makes Uhl's conception seem dubious.

3. It is unthinkable that an extensive alteration of the myoepicardiac mantle at the time which Uhl considered to be most probable could lead to an isolated myocardiac anomaly in the right ventricular region, quite apart from the fact that unilateral arrestation in the developement of the myocardium during the bilateral symmetrical phase is an absolutely untenable thesis.

Table 1. Compilation of all publications concerning Uhl's disease

| Author | No. of cases | Age | Sex | Cause of death | Diagnosis | Weight of heart (g) | Extent/localization of the pardement region in the right ventricle | Additional anomalies |
|---|---|---|---|---|---|---|---|---|
| Osler (1905)<br>Segall (1950) | 1 | Parchment heart | | | | | | |
| Uhl (1952) | 1 | 8 months | ♀ | cardiac failure | p.-m.e. | | inflow tract | |
| Castleman (1952) | 1 | 24 years | ♀ | cardiac failure | p.-m.e. | 300 | | |
| Novak (1957)<br>Arcilla (1961) | 1 | 5 months | ♂ | right cardiac failure | p.-m.e. | | whole free wall | patent for. ovale without shunt |
| Lepoix (1958) | 1 | 6 months | | cardiac failure | p.-m.e. | | | pulmonary atresia, absence of tric. valv |
| Gacul (1960) | 2 | 1 year<br>1 year | | cardiac failure | p.-m.e.<br>clininal | | | |
| Hasegawa (1963) | 1 | 6 years | ♀ | post Glenn-Op. | p.-m.e. | 160 | | patent for. ovale |
| Reeve (1964) | 1 | 47 years | ♀ | subarachnoid hemorrhage | p.-m.e. | 290 | $3 \times 7$ cm | patent for. ovale |
| Perrin (1965) | 2 | 10 months | ♂ | cardiac failure | p.-m.e. | | ant. free wall | atrial septal defect |
| (Taussig, 1960) | | 6 years | ♂ | post Glenn-Op. | p.-m.e. | | inflow tract | |
| Neimann (1965) | 1 | newborn | ♀ | cardiac failure | p.-m.e. | | whole free wall | pulmon. atresia, absence of tric. valves |
| Cumming (1965) | 1 | 7 months | ♀ | post Glenn-Op. | p.-m.e. | 48 | ant. free wall | patent for. ovale |
| Gould (1967) | 1 | 66 years | ♂ | chronic myeloid leukemia | p.-m.e. | 600 | $5 \times 5.5$ cm | |

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|---|---|---|---|---|---|---|---|---|
| Froment (1968) | 5 | 19 years<br>26 years<br>40 years<br>15 years<br>17 years | ♂<br>♂<br>♂<br>♂<br>♂ | cardiac failure<br>cardiac failure | p.-m.e.<br>p.-m.e.<br>clinical<br>clincial<br>clincial | 350<br>430 | | |
| Bansi (1968) | 1 | 68 years | ♂ | cardiac failure | p.-m.e. | 400 | | |
| Kinare (1969) | 1 | 5 years | ♀ | cardiac failure | p.-m.e. | 130 | whole free wall | |
| Montella (1969) | 1 | 38 years | ♀ | cardiac failure | p.-m.e. | | ant. free wall | |
| Matsuo (1969) | 1 | 5 days | ♀ | cardiac failure | p.-m.e. | | | |
| Matsuo (1969) | 1 | 5 days | ♀ | cardiac failure | p.-m.e. | | | pulmonary stenosis, patent for. ovale |
| Ohonogi (1969) | 1 | | | | | | | |
| Sugiura (1970) | 1 | 84 years | ♀ | pneumonia/ileus | p.-m.e. | 220 | outflow tract | |
| Zuberbuhler (1970) | 2 | 17 months<br>15 months | ♀<br>♂ | intra Glenn-Op. | p.-m.e.<br>clinical | | | |
| Tadei (1971) | 1 | 22 years | | cardiac failure | p.-m.e. | | | |
| Morand (1972) | 1 | 20 years | ♂ | | clinical | | | |
| Forssman (1972) | 1 | 47 years | ♂ | ventricular fibrillation | p.-m.e. | | | |
| Diaz (1973) | 1 | 4 months | ♂ | cardiac failure | p.-m.e. | | inflow tract | |
| Abe (1973) | 1 | | | | | | | |
| This case (1973) | 1 | 71 years | ♀ | left cardiac failure | p.-m.e. | 480 | inflow tract | disturbance in development of ostium-AV-dextrum |

p.-m.e. = post mortem examination

If one part of the embryologic cardiac system is retarded in growth, it is suppressed by the contralateral sections which continue in grow (Kl. Goerttler, 1958).

The external form of the myoepicardiac mantle determines the progress of the filaments of the blood stream, upon which again the shaping of the endocardiac structure and septa is dependent (Kl. Goerttler, 1955; Doerr, 1964).

In the damaging of the myocardium according Uhl's conception, therefore, there would appear as additional anomalies on the one hand alterations of the external form of the heart and on the other one would have to expect ventricular-septum-defects and deformation of the bulbus-truncus-region.

A theory which is to be acceptable in all its points and which intends to explain the morphogenesis of Uhl's disease must take basically two factors into consideration:

1. No anomalies can be found in the ventricular septal area or in the downwards bulbus-truncus-region.

2. The original hypothetical disturbance of the myocardium affects isolated right-ventricular areas; the left-ventricular myocardium is perfectly normal.

Accordingly it must be accepted as certain that the teratogenetic determination of Uhl's anomaly is to be place after ventricular septation has taken place, or in the last stages of this process, also after the vectorial bulbus turn has developed as normal. In order further to limit the determination time for the partial parchment heart, one must keep in mind that the formal principle of teratogenesis is not that of destruction of already formed structures, but of a retardation or an arrest of the normal process of structural formation (Büchner, 1952). The formation of a certain structure can however only be retarded in isolation by the surrounding formations when at a certain stage in its growth, in its mitotic rate, it dominates the neighboring zones, in other words, when the vulnerability at a particular time is limited to the area in question.

Towards the end of the 5th week of embryogenic developement together with the parallel orientation of both ventricles the right ventricle becomes conspicuous, the wall thickens noticeably until it is in excess of that of the left ventricle by a little. Even before birth and together with the decrease in width of the ductus arteriosus Botalli the left ventricle becomes somewhat stronger. Immediatly after birth the wallthickness of the right ventricle diminishes both absolutely and relatively (Kl. Goerttler, 1958).

Correspondingly we must expect the myocardium of the left ventricle to be more susceptible of damage until the end of the 5th week of embryonic developement, but form the 6th week on it is that of the right ventricle which is more vulnerable.

According to Kl. Goerttler (1956, 1957) the pathological processes in this phase are a matter more of degenerative than of inflammatory phenomena.

Therefore Uhl's disease according to the above-mentioned theory would be classified under differentatory disturbances of the heart in the tertiar period (cyematopathy system 3) (from the 2nd half of the 2nd month of developement) and would correlate in time with the determination of Ebstein's anomaly, Fallots' trilogy and the Lutembacher-Cossio syndrome.

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