The background of the cover is a high-magnification, blue-tinted micrograph of retinal tissue. It shows the intricate, wavy structure of the outer plexiform layer, with numerous small, dark, circular or oval-shaped cells (likely photoreceptors or Müller cells) interspersed among the fiber-like structures. The overall appearance is that of a complex, layered biological structure.

Stereoatlas of Ophthalmic Pathology

Anatomy and Pathology of the
Peripheral Fundus (Fundus extremus)

A bequest of Basil Dalcker
Former Professor of Ophthalmic Pathology in Basel

Editors
Peter Meyer, Basel
Karin U. Löffler, Bonn

KARGER

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246 stereoscopic figures, 240 in color and 1 schema,
includes stereo glasses and a CD-ROM, 2006

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Only thanks to the ample financial support of the La Roche Foundation Basel and the Freiwillige Akademische Gesellschaft (FAG, Voluntary Academic Society) in Basel is it possible to produce the stereoatlas in its present and generously illustrated form.

Special thanks also go to my family for their patience and understanding. Finally, I would like to thank Karger Publishers and its staff for their untiring assistance in the preparation of this atlas.

Peter Meyer, MD

Foreword

by Josef Flammer, MD, Professor,
Department of Ophthalmology, University of Basel
and Michael J. Mihatsch, MD, Professor,
Department of Pathology, University of Basel

Ophthalmologists are in the privileged position to ‘directly see’ some of the ocular diseases. However, only certain aspects of eye diseases can be appreciated employing biomicroscopic techniques. Often additional very important findings only become apparent after careful gross and histologic examination of surgical ophthalmic resection specimens. Close clinicopathologic correlation not only provides important clues for direct patient management but also gives insight into the pathophysiology of the underlying disease processes. Such correlations help to ‘ask’ scientific questions and to target research projects that further improve our understanding of ocular diseases and patient management. Prof. Basil Daicker was a pioneer in this field.

Prof. Basil Daicker had a unique personality: he combined the expertise of a clinical ophthalmologist with the skills of an ocular pathologist, the attitude of an artist and the ‘fears’ of a perfectionist. The latter characteristic prevented him from publishing his outstanding collection of more than 10,000 stereophotographs and corresponding histopathologic images. He always felt that his work needed further improvement. Prof. Basil Daicker, who unfortunately passed away shortly after his retirement, left his impressive collection of pictures to his successor Dr. Peter Meyer. Dr. Meyer took it upon himself to review Prof. Daicker’s collection and to select representative images for publication. Thanks to Dr. Meyer’s effort, Prof. Basil Daicker’s work will survive as an exquisite tool for studying ophthalmological pathology.

We hope that the readers, both young trainees and experienced ophthalmologists, will benefit from working with this atlas that will in our opinion become one of the standard books in ophthalmology.

Foreword

by Heinrich Witschel, MD,
former Head of the Department of Ophthalmology
University Hospital Freiburg i. Br., Germany

I first met Basil Daicker in the early 1970s. We, that is the residents of the University Eye Hospital Freiburg im Breisgau, had come to Basel to listen to one of the famous afternoon lectures he used to give for residents, staff members and practicing ophthalmologists. I vividly remember how fascinated we were by the wonderful and most instructive transparencies, most of them in stereo, with the help of which Daicker introduced us to the ‘miracles’ of the peripheral fundus of the human eye, an area which had only recently been made accessible to the clinician by the new tools of the three-mirror contact lens and of indirect ophthalmoscopy with indentation.

For me this experience was the starting point for my own scientific approach to the peripheral retina and its diseases. Basil and I became friends later on. As often as possible I went to Basel to learn from his superb preparation technique both macroscopic and microscopic, and though I succeeded to a certain degree, I never reached or even approached his skills and his patience when he was meticulously preparing and photographing the specimens with the simple instruments available in those years.

At that time the tradition of ophthalmic pathology, most of which had been lost during the Second World War, was enjoying some kind of renaissance. This revival was substantially supported by the foundation of the European Ophthalmic Pathology Society in 1962 and the Society of German-Speaking Ophthalmopathologists in 1972. Basil Daicker was a member of both societies and he made many valuable contributions to each of them. To my understanding Basil Daicker was a prominent exponent of the unfortunately decreasing number of scientists whose way of doing ophthalmic pathology was in keeping with the purpose

it was originally meant for, namely in the first instance to closely correlate clinical findings with macroscopic and microscopic histopathology and only secondly to deal with extremely rare tumors or similar lesions.

Ophthalmic pathology as a speciality developed during the 19th century paralleling the evolution of general pathology. It has always kept pace with the latter and has made use of its modern techniques, but it has never been a branch of it. With very few exceptions the general pathologists were not interested in the structure and the diseases of the eye. Thus the founders of ophthalmic pathology were physicians practicing clinical ophthalmology. The most prominent among them were Otto Becker (1828–1890) in Heidelberg, Theodor Leber (1840–1917) in Göttingen and Heidelberg, Julius Michel (1843–1911) in Würzburg and above all Ernst Fuchs (1851–1930) in Vienna, whose contributions to our knowledge of the histopathology of the eye cannot be overestimated. These men and their successors took the questions arising during their daily clinical work immediately to their laboratories.

The method of directly correlating the observations the clinician makes with the help of the already considerably magnifying slit lamp and the ophthalmoscope with the macroscopic and microscopic findings in the laboratory has substantially contributed to the permanent progress in clinical ophthalmology, including ophthalmic surgery. As a consequence today most ophthalmic pathology laboratories are run by clinicians with specialist training in pathology, at least in continental Europe.

Basil Daicker has left us with a treasure of more than 10,000 excellent and instructive transparencies to bear witness to his profound knowledge of ophthalmic

pathology and his outstanding skills in preparing and photographing ophthalmic specimens. This collection must not be allowed to get lost. Thus we, that is the 'community of ophthalmic pathologists', are very grateful to Dr. Peter Meyer for editing this book and making a first series of pictures accessible to the public. We hope that this volume will be only the first of a larger series opening more of the valuable collection to future generations of ophthalmologists.

Preface

Hardly any subarea of ophthalmic pathology is as multifaceted as the retinociliary periphery of the ocular fundus or – in Professor Basil Daicker’s words – the ‘fundus extremus’. Be it degenerative changes, inflammatory processes, tumors or trauma to this region, the opportunity to evaluate the clinical and morphological picture at the same time has up to now had the most crucial impact on our patients. Ophthalmic pathology provides the basis for a clinical diagnosis and its appropriate therapy, in particular for the ophthalmic surgeon, and thus represents an important tool of quality control.

In his 30 years as an ophthalmic pathologist at the Department of Ophthalmology, University of Basel, Switzerland, Professor Daicker contributed substantially to our actual understanding of ocular disease, especially regarding the area of the fundus extremus. In a large number of ocular specimens, using special investigation techniques, he analyzed the fine anatomical structure and its variations as well as a multitude of frequent and rare pathological findings. He documented his observations in a worldwide unique ophthalmopathological collection and, between 1965 and 1995, assembled and filed more than 10,000 stereoscopic slides of interesting and uncommon diseases.

Here, in memory of Basil Daicker, part of this inimitable collection is for the first time made available to the public. We have for the time being restricted ourselves to the fundus extremus, with Daicker’s monograph on the anatomy and pathology of the human retinociliary fundus periphery (published in 1972 by Karger, Basel) serving as a template. Similar to his previous publication, we subdivided the stereoscopic atlas into seven chapters with a short summary at the beginning of each. The extremely high standard and to some extent matchless stereoscopic pictures represent the focus of each chapter. Based on Professor

Daicker's autopsy and histology reports, we adopted his observations and described the various disease patterns accordingly.

It was our wish to make these disease patterns compiled by Daicker available to the ophthalmic pathologist as well as to the clinician. Due to the stereoscopic effect, they appear particularly lifelike and memorable. The pictures can be consulted by the ophthalmologist for diagnostic purposes, while for the layperson, they provide an insight into the beautiful microcosmos of the inner eye.

Peter Meyer, MD

Portrait



Basil Daicker

Professor Daicker was born in Basel, Switzerland, in 1932. After studying medicine in Basel until 1958 he worked as a Resident in the Department of Ophthalmology at the University of Basel, where he obtained his doctorate in medicine in 1962. In 1965, Basil Daicker became Head of the Division of Ophthalmic Pathology.

In 1970 he obtained his 'venia docendi' at the University of Basel with a widely recognized monograph on the morphology and pathologic changes of the peripheral fundus. For his various investigations involving retinal diseases and the role of fundus diseases which were considered to be predisposing to the development of retinal detachment, he was awarded the Alfred Vogt Prize (1970) and the Graefe Prize (1979). He brought Ophthalmic Pathology to a high scientific level and was an excellent teacher of clinicians and scientists.

Since 1972 he was a regular member of EOPS (European Ophthalmic Pathology Society). He died in spring 1996, only a few month after his retirement. Posthumously, he was awarded an honorary membership by the Swiss Ophthalmological Society (SOG) in Geneva.



Normal Anatomy

Introduction

Normal Anatomy of the Peripheral Fundus (‘Fundus extremus’)

The retinociliary periphery of the ocular fundus was called ‘fundus extremus’ by Daicker. Anatomically, this is the ring-shaped region of the inner ocular layers, extending from the ora serrata over the pars plana of the ciliary body up to the pars plicata corporis ciliaris (see schema on p. 3).

The ora serrata in particular often shows individual differences. Various irregularities have been grouped into three characteristic subtypes [1, pp. 29–38]:

(1) Lissora, Spiculae of the Ora

Lissora describes the presence of biomicroscopically visible little rudimentary teeth around the whole circumference. Spiculae are extremely fine filiform extensions reaching from the retinal edge anteriorly into the vitreous over the ora.

(2) Ora serrata in the Narrower Sense,
Ora Teeth

The teeth-like morphology is already visible macroscopically and occurs primarily in the nasal part of the globe. The number of teeth varies individually.

(3) Cristae ciliares

Markedly longer ora spikes extend as a narrow band meridionally across the whole pars plana up until the area located between two ciliary processes. They are ridge-like excrescences of the orbiculus.

Pars plana

The pars plana corporis ciliaris (orbiculus ciliaris) extends as a ring-shaped zone between the ora serrata and the posterior border of the pars plicata corporis ciliaris. The orbiculus is broader temporally than on the nasal side. Anterior to the ora is the preretina, which while narrow on the nasal side has a broad lighter zone on the temporal side. In this area the ciliary epithelium is taller. Its spiky anterior border is called preora (schema below of fig. 1). Due to topographical peculiarities, especially of the zonules and the vitreous body, the region of the preretina plays a particularly important role in pathological processes of this area.



Zonule

The zonule, zonular fibers, or zonule of Zinn refer to a system of fibers which becomes more defined at the anterior pars plana, between the ciliary processes and their lens attachment. Two major systems of long zonules are closely related to the orbiculus and the adjacent vitreous: the anterior (outer) orbiculocapsular and the posterior (inner) orbiculocapsular system [1, pp. 81–90].

Fibers of the anterior orbiculocapsular system originate from fibrils emanating from the basal lamina of the ciliary epithelium in the orbiculus region. They merge to form thick fibers and extend through the valleys between the ciliary processes up to their insertion into the anterior lens capsule. The most posterior origin of this fiber system coincides with the anterior border of the preretina or the preora. The posterior orbiculocapsular system comprises less robust fibers above the orbiculus. Usually they enter in the posterior aspect of the zonular attachment to the lens. Most of the fibrils originate from the most anterior inner surface of the vitreous base. Further insertion points are located within the vitreous scaffold of the vitreous base in the ciliary as well as in the retinal part and also in the paracortical vitreous in front of the tractus Retzii.

The circular fibers form three fine bands [2, pp. 15–16]:

- (1) One located most posteriorly above the mid pars plana (ligamentum medianum of the pars plana, posterior orbicular circular band, fig. 2 and 3).
- (2) One located above the posterior third of the corona ciliaris (ligamentum coronarium, anterior orbicular circular band of membrana hyaloidea, band of Salzmann, fig. 12).
- (3) One located anterior just behind the edge of the lens (fasciculus retrolentalis).

Both posterior zonular bands are connected to the ciliary body by fibers. In addition, there are numerous finer fibrillar systems, connecting the

main fibers to the ciliary body, which bridge adjoining ciliary processes or are penetrated just above the vitreous.

Vitreous Body

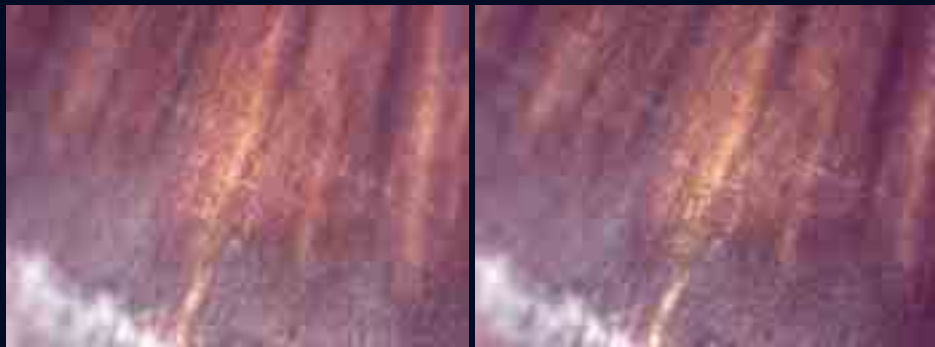
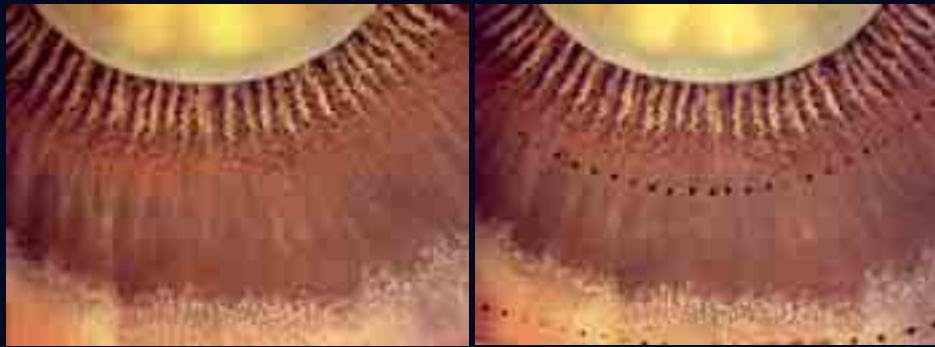
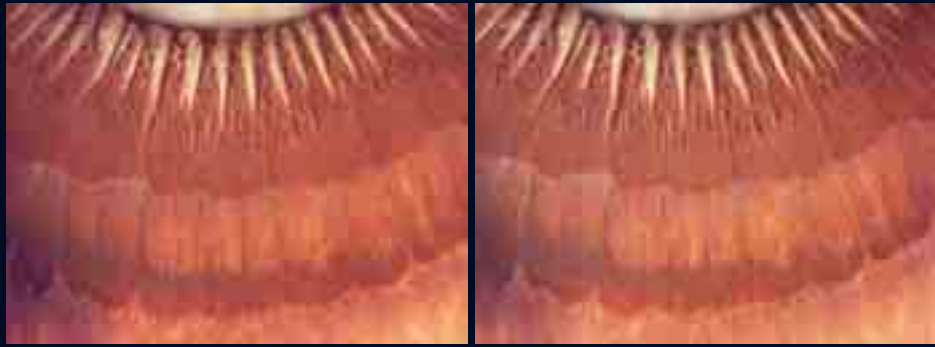
The anatomical and topographic conditions of the vitreous in the area of the ora serrata play a clinically significant role with regard to the mechanisms leading to retinal detachment. There is a ring of laminar adhesion (the so-called pars retinalis of the vitreous base) between the peripheral retina and the vitreous cortex that enlarges towards the equator with age. The area of the most powerful vitreous attachment is located anterior to the ora and extends up to the posterior insertion point of the zonules at the ciliary epithelium of the orbiculus [3]. This area is called the pars ciliaris of the basis vitrei and according to Daicker corresponds to the area of the preretina [1, pp. 69–71]. The anterior vitreous surface is defined by a condensation of vitreous fibrils (membrana hyaloidea anterior = anterior limiting membrane of the vitreous). When detached from the posterior lens surface (at the band of Wieger), it extends in a ring-like convex fashion over the posterior zonules and the pars plicata corporis ciliaris. Above the posterior pars plicata the hyaloid membrane merges with the band of Salzmann. Between the orbicular part of the hyaloid membrane and the ciliary epithelium of the orbiculus there is a ring-shaped space approximately triangular when viewed as a cross section. This orbicular space is also called the space of Garnier after the person who first described it and is filled with aqueous fluid (fig. 14).

1 **Pars plicata, pars plana (orbiculus ciliaris) and ora:** Ciliary processes are slim and have even surfaces. The border between the anterior part and the posterior pars plana is the linea serrata of the pars plana (preora). The anterior pars plana is dark, the posterior pars plana appears bright and is called the preretina. It is the area where the ciliary epithelium is taller. A broad anulus pigmentosus ciliaris (marked area) can be seen.

2 **Pars plicata, pars plana (orbiculus ciliaris) and ora in an adult:** In the midst of the pars plana circular fibers of the zonular network form the ligamentum medianum (midline) across the ciliary epithelium of the pre-ora region. The anterior hyaloid membrane is connected to the ciliary body by fibers of the ligamentum medianum.

3 **Detail of figure 2:** Midline and side line can be seen.

4 **Section of the anterior calotte (equatorial opening):** Anterior to the ora there is a broad fairly light-colored band with a toothed border anteriorly, corresponding to the taller ciliary epithelium, the preretina. Originating from the preora, small meridional striae epitheliales can be seen adjacent to the well-discernable midline.



5 **Ora and ciliary body, 28th week of pregnancy:** Orbiculus is still small; variably prominent ora indentations can be seen.

6 **Ora and ciliary body in a fetus, 28th week of pregnancy:** Narrow pars plana with variable indentations (teeth) of the ora serrata. Striae ciliares epitheliales can be seen: thin meridional lines of light pigmentation, originating from ora teeth and running as narrow bands across the orbiculus ciliaris into a valley between two ciliary processes where they merge furrow-like with their surroundings.

7 **Adult nasal ora:** Spiculae along teeth and bays. Early cystoid retinal degeneration.

8 **Ciliary processes with zonular fibers in an adult:** The anterior and posterior (ciliocapsular) leaves pass from the corona ciliaris to the lens. They are attached to the zonular lamella, anteriorly at about 1 mm, posteriorly at a distance of 1.5 mm from the equator.



9 **Posterior insertion site of the zonules:** The insertion site at the posterior zonular lamella of the lens is clearly visible.

10 **Same situation as shown in figure 9:** Lateral view.

11 **Ciliary processes with zonular fibers in a 4-year-old child:** Beneath the zonules the back of the iris is clearly visible.

12 **Anterior orbicular ciliary band of the anterior hyaloid membrane, the so-called band of Salzmann:** Firm collagen fiber bundles are merging concentrically to the equator of the lens with the hyaloid membrane, often appearing as multiple small folds. This ring-shaped band is called band of Salzmann after the person who first described it.

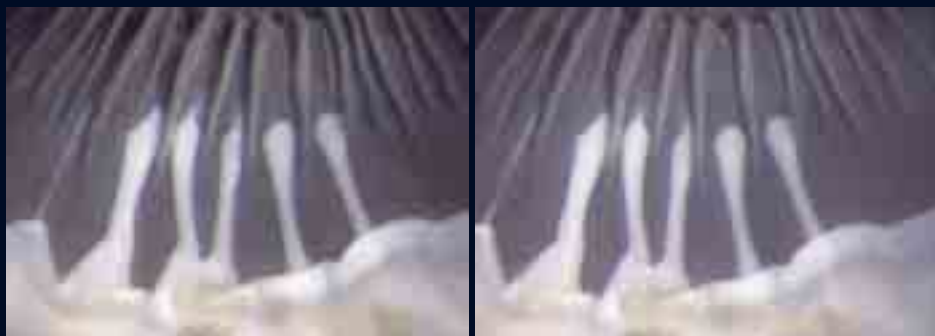
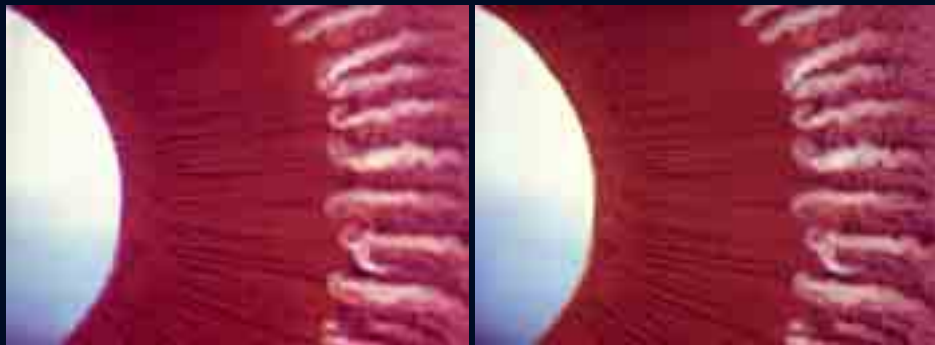
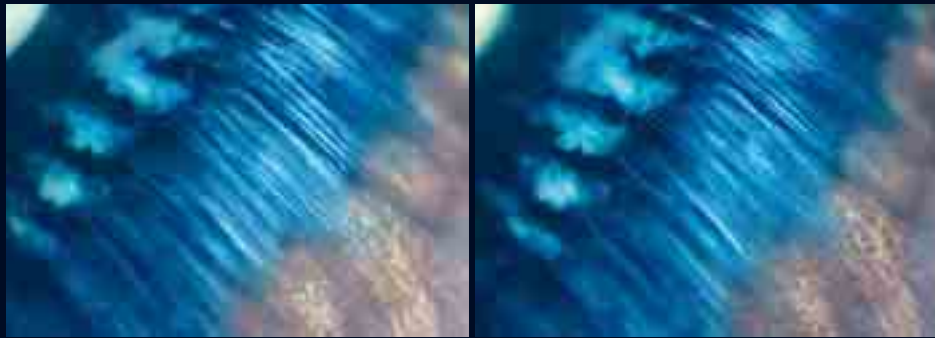
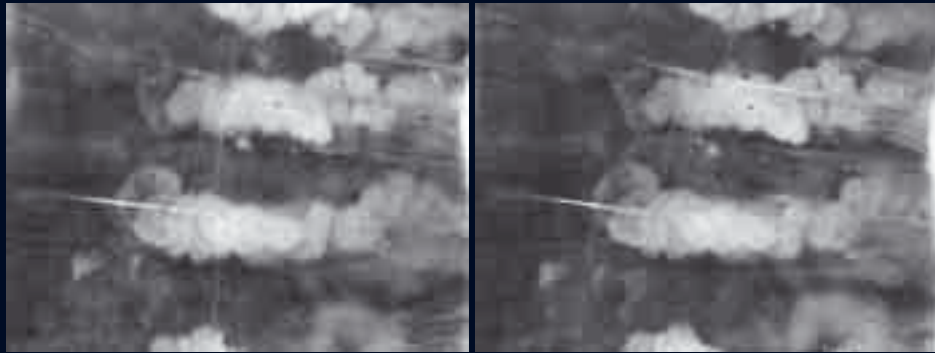


13 **Same case as shown in figure 12:** Insertion of orbiculovitreal ligaments into the anterior ciliary band.

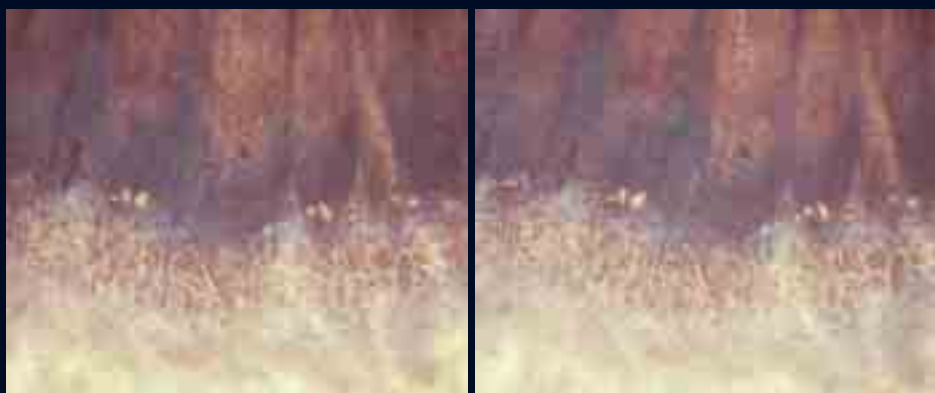
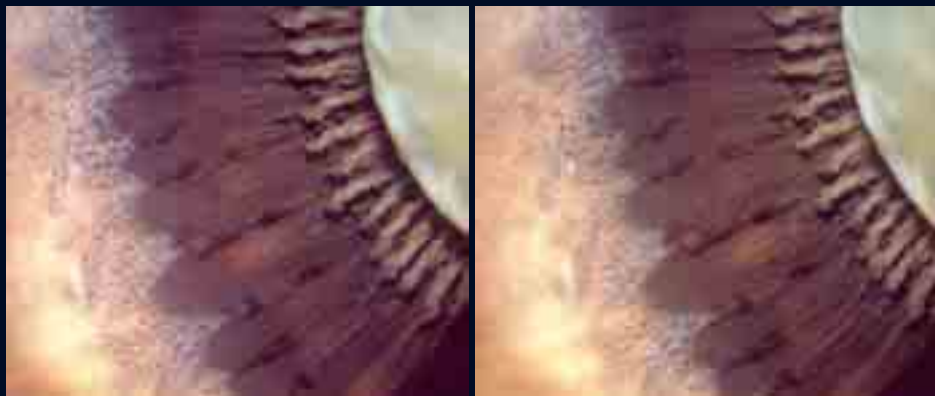
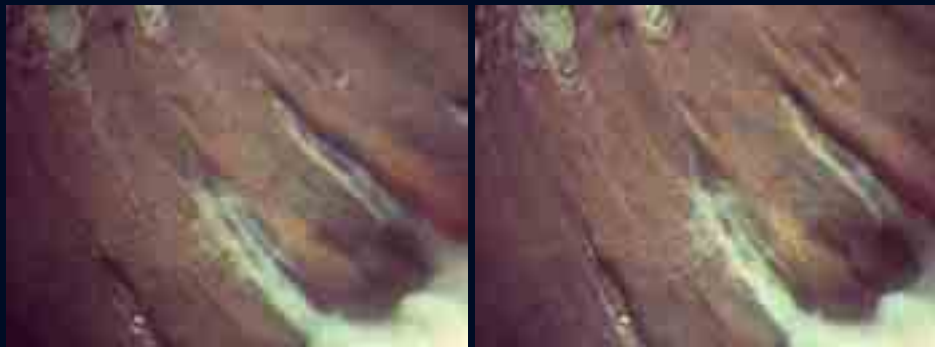
14 **Posterior chamber:** By instillation of dye (Alcian blue 1%) into the posterior chamber the space of Garnier is well demarcated and is, like the zonules, clearly visible.

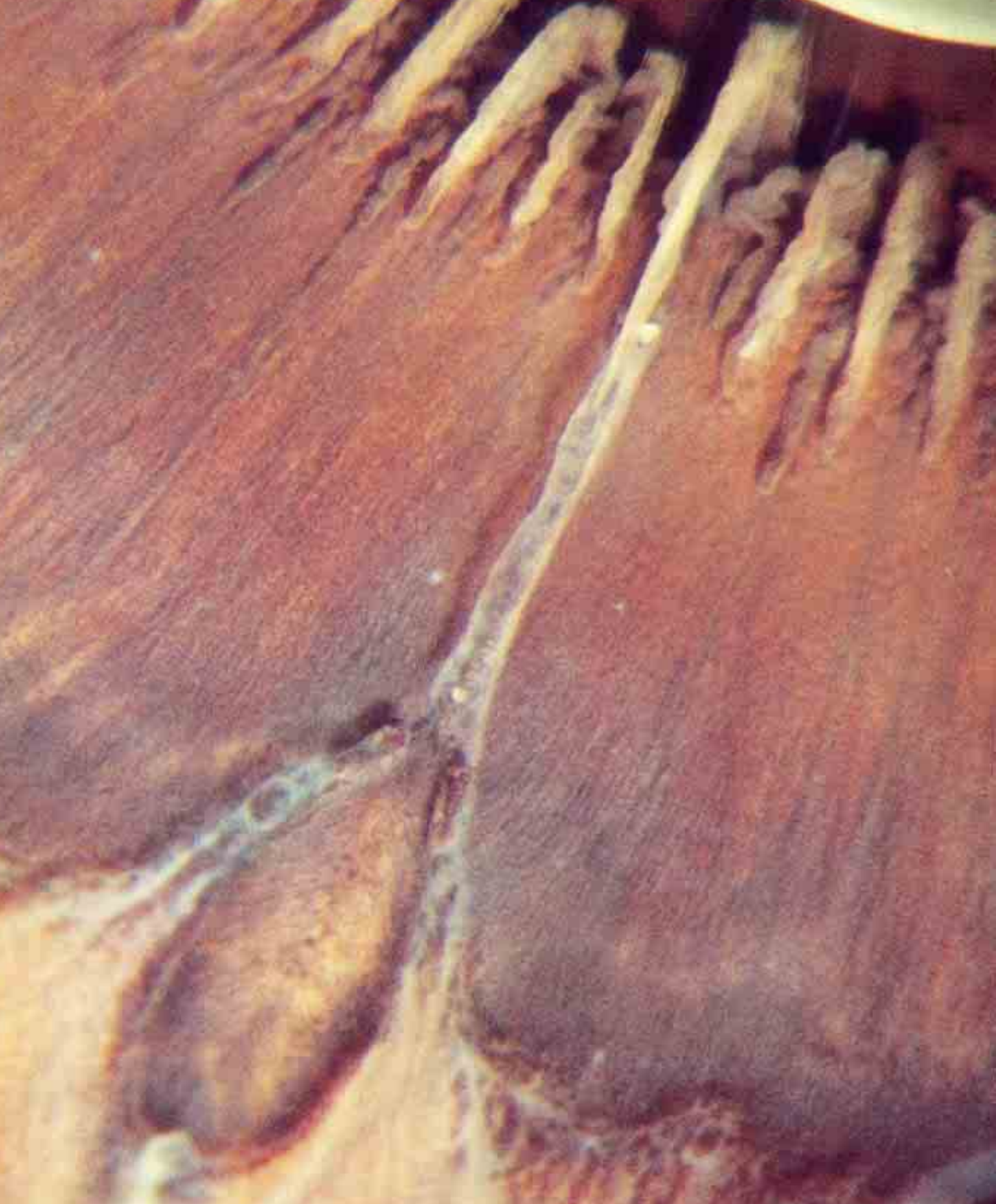
15 **Posterior iris surface:** After removal of the lens, the delicately lamellated relief of the posterior iris surface can be seen.

16 **Multiple cristae ciliares retinales in a newborn:** Slender ridges with a bulbous swelling anteriorly extend over the orbiculus, and a Lange fold covers the ora.



- 17 Cristae ciliares (pars plana ridges) in an adult, originating from long ora teeth on the nasal side, are shown.
- 18 Same case as shown in figure 17. Cristae ciliares with pinnacle-like proliferation of the ciliary epithelium.
- 19 Multiple pigmented pars plana ridges nasally, adjacent to an area of pronounced cystoid retinal degeneration.
- 20 **Vessels of Sattler:** Tight network formed by capillaries in the region of the ora. This particular vascular system is located between Bruch's membrane and the pigment epithelium and further anteriorly within the interlamellar fibrous tissue of the pars plana between the elastic lamina and the basal lamina of the pigment epithelium. These vessels develop after the age of 50 [4, 5].





Congenital Malformations

Introduction

The peripheral fundus shows an extremely high degree of individual variations. Micromalformations such as ora teeth extending anteriorly far beyond the orbiculus, pars plana ridges and ciliary processes of the third order are so frequent that Daicker regarded them as normal. These findings are, therefore, dealt with in the chapter of normal anatomy.

The following selection of some more unusual micromalformations will be presented at the beginning of this chapter: pathological ora bay (fig. 1), Y-shaped pars plana ridge (fig. 3) and pars plana island with malformed ora tooth (fig. 4).

While these variations are usually insignificant, there are, nevertheless, and not infrequently, also micromalformations that should be regarded as pathological since they can become clinically relevant. They are called either Schepens' folds after the person who first described them or meridional folds, reflecting their location. Daicker found them in about 12% of autopsy eyes. They are granular meridional protrusions on top of ora teeth that extend anteriorly, sometimes up into the anterior third of the orbiculus, and by about the same length onto the retina posteriorly towards the equator. Histologically they are composed of strands, tubuli and convolutes of epithelial cells above a gliotic and atrophic retina. Even in fetal eyes one can find the beginning of meridional folds (fig. 8).

Spencer et al. [6] described a special type of meridional ridge (fold) which he called the meridional complex. These meridional ridges are exceptionally long, extending anteriorly into the ciliary process. According to studies of Eisner

[2, pp. 57–58], meridional ridges are not dangerous so that prophylactic photocoagulation is unnecessary.

The most frequent malformation of the peripheral fundus is the rosette (clinically present in 27–30%, see Trantas [7], and in autopsy eyes in 43%, see Daicker [1, p. 107]). A rosette represents an epiretinal epithelial proliferation in the configuration of warts, lappets or more laminar entities (which the Americans therefore call granular patches). In addition, excessively long worm-like structures can occasionally be observed (fig. 21). Free-floating epithelial conglomerates (free rosettes) can sometimes be seen in the peripheral vitreous close to the retina.

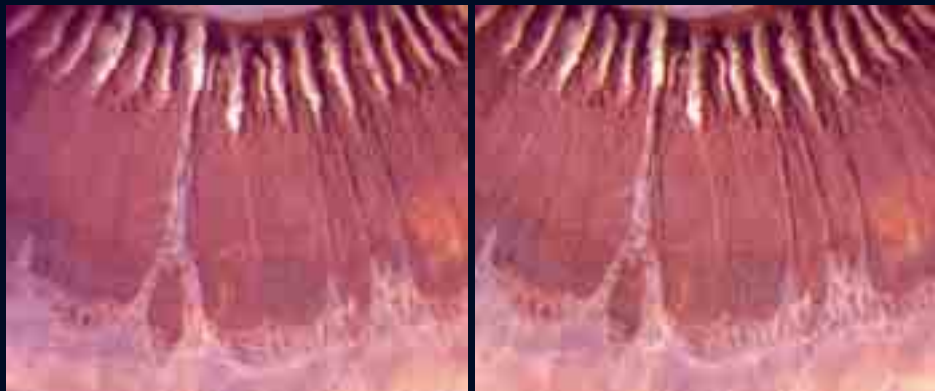
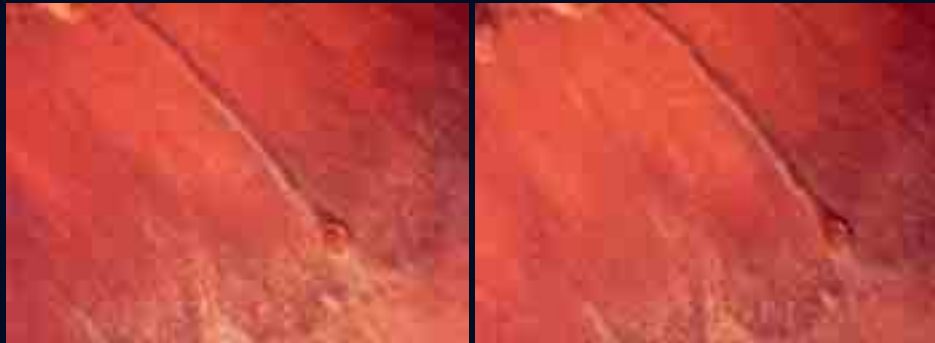
The final part of this chapter will present grossly dysplastic changes of the peripheral fundus. They are nearly always combined with other malformations of the globe and are usually found in microphthalmic eyes. Coloboma of the ciliary body represents the most serious inhibitory malformation in this area and is typically found in the lower eye cup.

1 Wide and deep pathological ora bay temporally surrounded by pronounced cystoid degeneration of the peripheral retina.

2 Pars plana ridge with a druse near the ora serrata.

3 **Y-shaped pars plana ridge:** Two neighboring ora teeth converge into a ridge which fuses anteriorly with a ciliary villus. Adjacent, there are three additional pars plana ridges.

4 Large pars plana island in the most peripheral retina with a rosette posteriorly and a Y-shaped pars plana ridge with drusen anteriorly.



5 **Temporal ora:** Double ridge with drusen embracing a pars plana island anterior to an area of microcystoid degeneration.

6 Two meridional folds extending from the most peripheral retina (with cystoid degeneration) towards the middle and anterior orbiculus. They are composed of rather dense tissue alternating with more transparent tissue.

7 **7-year-old child:** Two meridional folds originating in a slightly dysplastic ora; the peripheral retina shows a wart-like rosette.

8 Meridional complex in a newborn originating from an area of early cystoid degeneration in the peripheral retina and radiating outwards to the ciliary villi.



- 9 Four meridional complexes between ora and pars plicata.
- 10 Two meridional complexes from case 9, originating from two ora teeth. The composition of individual dense as well as translucent portions is obvious.
- 11 Distinct meridional complex with equatorial degeneration at the posterior border of the ridge and adjacent peripheral cystoid degeneration.
- 12 Extremely large meridional complex with multiple frayed rosettes at its posterior border in the peripheral retina. A pars plana ridge is seen at the top.



- 13 Merdional complex with pars plana island and meridional fold anteriorly, originating in a retina with cystoid degeneration.
- 14 Zonule inserted into a meridional complex with a pars plana island at the posterior border. The orbicular space (posterior chamber) is stained in blue.
- 15 Multiple small filiform rosettes in the region of the ora.
- 16 Filiform floating rosettes near the ora, some of a rather bizarre appearance.



- 17 Free-floating and pedunculated pear-shaped pre-retinal rosettes anterior to the ora. Peripheral cystoid degeneration.
- 18 Behind an ora tooth there is a pars plana island with a wart-like rosette originating from its posterior border. Meridional folds can be seen on both sides.
- 19 Multiple free rosettes of variable size composed of condensed cellular elements, lying over the pars plicata.
- 20 Nonattached rosette over the pars plicata with part of a pars plana cyst behind it.

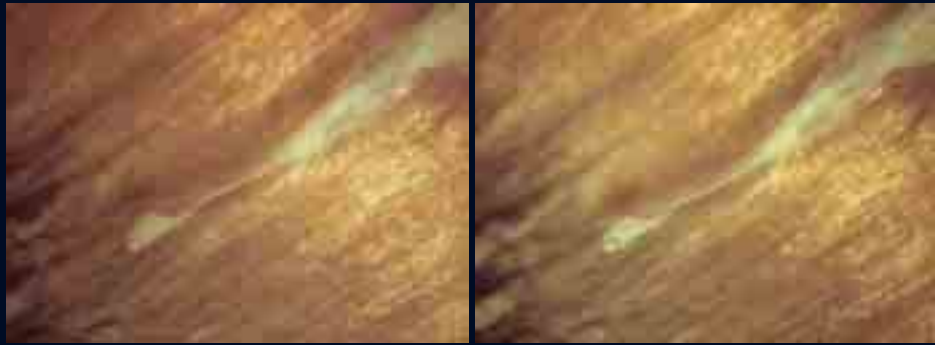


21 Pars plana ridge with worm-shaped pedunculated rosette.

22 **Fetus (34th week of gestation) with multiple malformations:** There are numerous rosettes on the peripheral retina, and in addition the fundus is covered by intraretinal hemorrhage.

23 Prominent wart-shaped retinal rosette with adjacent cystoid retinal degeneration.

24 Worm-shaped rosette over the ora serrata, merging with three retinal holes posteriorly. Note the neighboring cystoid retinal degeneration.



25 Behind the ora, there is a broad pedunculated rosette with a degenerative partial retinal hole at its base. The posterior border of the vitreous base is located on the same level as this rosette; vitreous condensations insert at its knoll. Note marked cystoid degeneration of the adjacent retina.

26 Pedunculated wart-like retinal rosette with an atrophic hole at its base surrounded by marked cystoid degeneration.

27 Worm-like giant rosette, extending from an area of gliotic/atrophic retina with a degenerative hole onto the pars plana.

28 Intraretinal worm-like rosette over a chorioretinal scar.

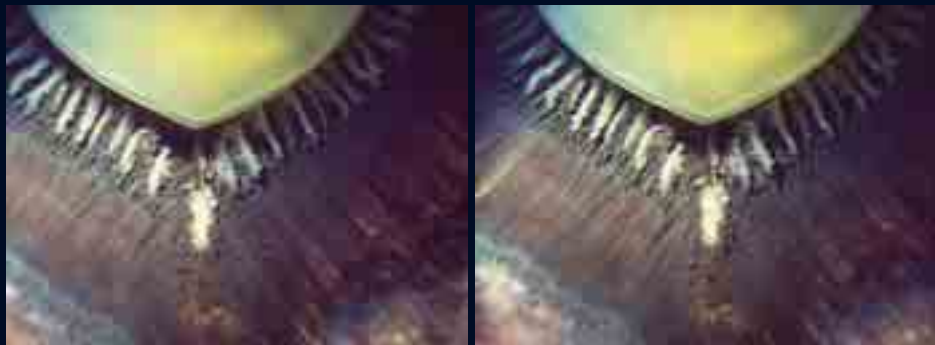


29 Long cone-shaped rosette of the peripheral retina with pigmentation at the base and zonular insertion on top.

30 Retinal atrophy with chorioretinal scar and small rosette in the center.

31 Coloboma of the iris and ciliary body can be seen below. The cataractous lens is indented and the zonules are missing in the area of the coloboma. A long vermiform rosette extends from the lower border of the coloboma into the vitreous. A meridional fold can be seen on the opposite side.

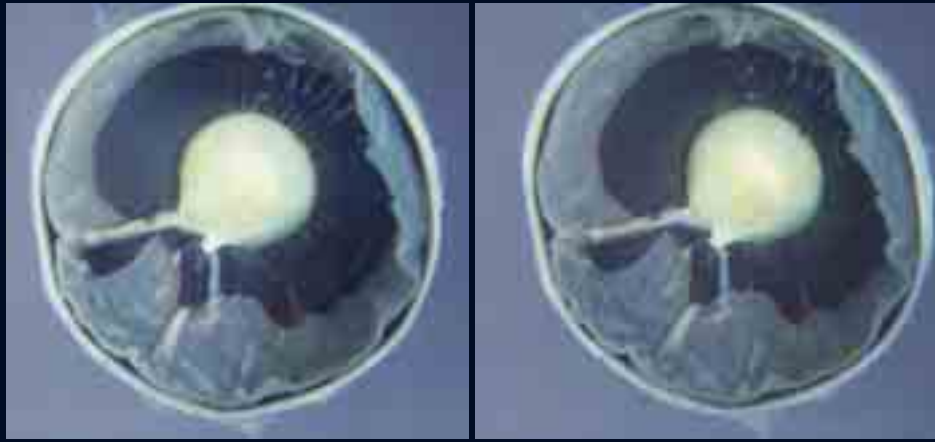
32 **Inferior coloboma of iris, ciliary body and lens:** The lens appears pointed below as does the pars plicata, and the most inferior ciliary process is markedly hyperplastic. The lower pars plana is broadened.

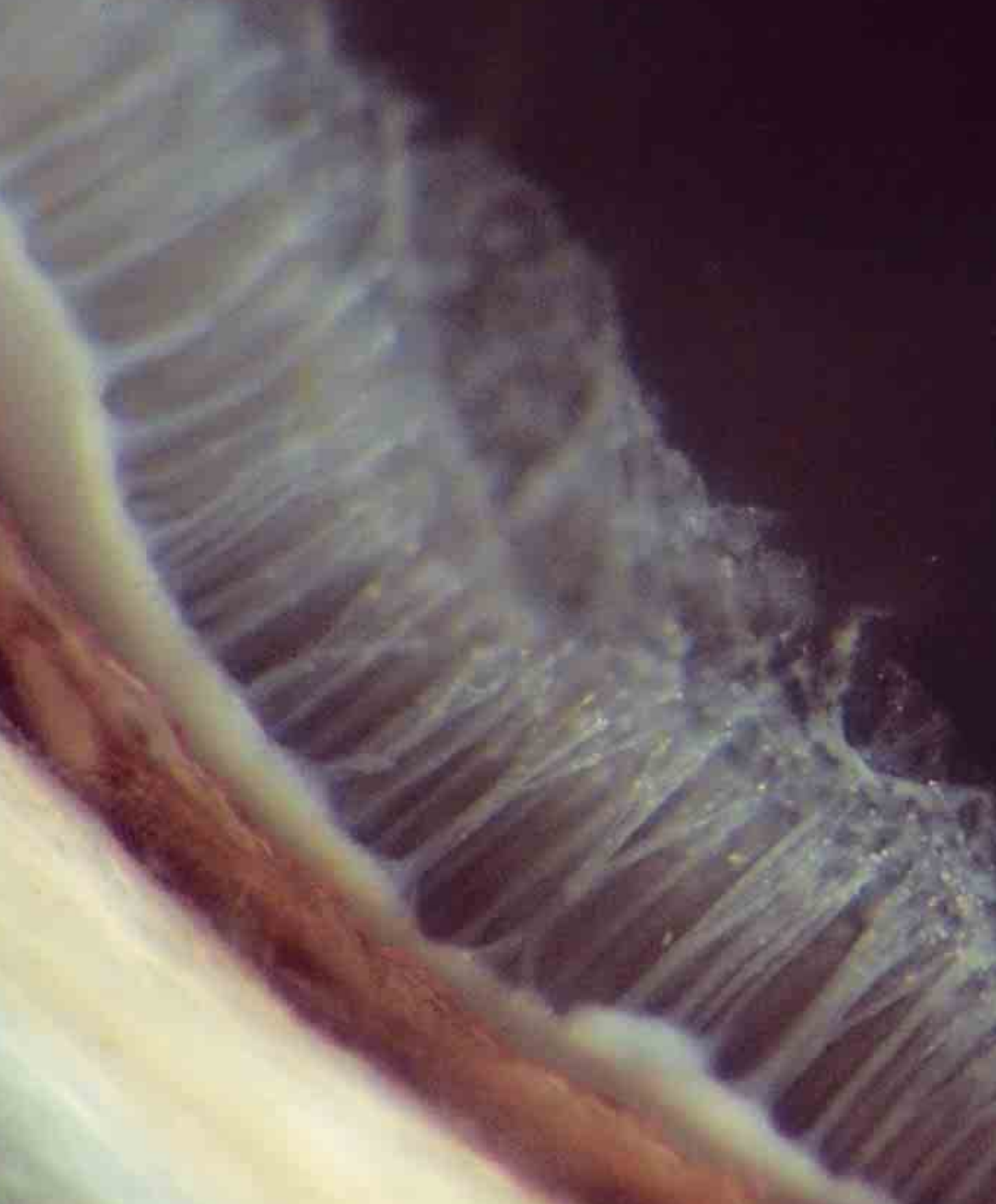


33 **Coloboma of the ciliary body:** Two retinal projections extend towards the posterior lens surface, and in between the retinal tissue reaches the ciliary processes. Ora, pars plana and pars plicata appear partly normal. On the opposite pars plana cilio-retinal crests can be seen.

34 **Detail of figure 33:** Projections of retina, ciliary epithelium and retinal pigment epithelium extend towards the posterior lens surface; the retinal strand terminates just anterior to it.

35 **Dysmaturity, 38th week of gestation, microphthalmos:** There is an isolated round coloboma of the ciliary body in the lower temporal quadrant. A worm-like rosette arises from the peripheral retina and extends over the pars plana up to the ciliary processes. In the temporal upper quadrant multiple retinal veils can be seen, reaching the posterior iris surface anteriorly. All of the upper nasal peripheral retina, the ciliary body, and the upper lens merge in whitish callous scar tissue





Degenerative Lesions

Introduction

Cystoid Degeneration of the Peripheral Retina

Cystoid retinal degeneration, also called Blesig-Iwanoff cysts, is the most well-known alteration of the peripheral fundus (fundus extremus). Already starting early in life, it is present in almost all adults and is absent only in eyes with high myopia or primarily atrophic retina. Typically, the cystoid degeneration starts with a status lacunaris in the outer plexiform layer of the peripheral retina, commencing at the ora and progressing towards the equator. The cystoid spaces enlarge with time and merge until they form a kind of portico. The pillars are formed by residual glia, and the cystoid spaces contain hyaluronic acid. The extent of the area involved is usually symmetrical, and more pronounced at the temporal side indicating a genetic predisposition.

Retinoschisis and Peripheral Retinal Cysts

Advanced cystoid degeneration can progress to retinoschisis. The glial pillars between the 'bottom' and the 'top' of the retina have partially or completely disappeared. Thus, two distinct sheets are formed representing the outer- and innermost retinal tissue. It occurs in a relatively flat form (retinoschisis). By further separation and ballooning in such an area, the development of a peripheral retinal cyst (in fact a pseudocyst) can be observed.

Edema and Pseudocystic Degeneration of the Preretinal Ciliary Epithelium

With increasing age, the preretinal ciliary epithelium anterior to the ora can become thickened and less transparent. There is thinning and elongation of the preretinal epithelium. In between, there are split-shaped and pseudocystic cavities (pseudocystic degeneration). In more advanced stages, the epithelium becomes multilayered.

Cystic Elevation of the Ciliary Epithelium (Pars plana Cysts)

Pars plana cysts (in fact pseudocysts) are a localized separation of the nonpigmented ciliary epithelium and the pigment epithelium. They are mostly found in the temporal pars plana, are mostly transparent and occasionally discretely pigmented. Their size ranges between minute button-like structures and a maximum diameter across the whole width of the orbiculus from the ora up to the posterior border of the pars plicata. In the mid-orbiculus, macrocysts can enlarge. Towards the ora, they taper off in a pear-shaped configuration. Daicker found pars plana cysts in about 25% of all postmortem eyes [1, pp. 229–237].

Degeneration of the Peripheral Retinal Pigment Epithelium

With increasing age the pigment epithelium between the ora and the equator changes into a band-shaped pattern with a mottled and usually darker appearance. Depending on locally predominant alterations two different groups can be distinguished: the macular and the linear degenera-

tion. Macular degeneration embraces the cobblestone degeneration, pigment epithelial alterations under lattice degeneration of the retina as well as the peripheral drusenoid fundus. Cobblestone degeneration is located mostly in the lower temporal periphery between the ora and the equator, rarely extending onto the pars plana, and consists of roundish either solitary, grouped or often confluent spots with sharp edges. Defects of the pigment epithelium, characterizing these spots, allow an unobscured view of the underlying choroidal vasculature. In larger spots, the overlying retina is still firmly attached. The linear degeneration is characterized by linear defects or proliferations of the retinal pigment epithelium. Daicker described three types of senile linear degeneration in contrast to traumatic linear changes: the *état craquelé*, the *degeneratio linearis vasculosa* and the *degeneratio reticularis* [8].

When viewed from above, the *état craquelé* appears as if lines, behind and mostly parallel to the ora, have broken up the mottled peripheral pigment epithelium. In small areas within the post-ora circumference patterns of vascular linear degeneration are seen especially with increasing age. Here the pigment epithelium is crossed by fine light-colored lines of a constant diameter. Frequently they form concave arches towards the ora, branching out into thinner irregularly running lines. Illuminated from the side, these lines appear somewhat elevated and wormlike. Histologically, newly formed vessels are found on the inner aspect of Bruch's membrane. In reticular degeneration the pigment epithelium forms hyperpigmented

lines, arranged in a polygonal meshwork. Along these lines numerous small hyaline drusen are present. Daicker observed this alteration behind the *état craquelé* going as far as the equator.

Degenerative Deposits on the Pars plana

In advanced cases of pseudoexfoliation syndrome, Busacca deposits are found not only along the lining of the posterior chamber but also on the pars plana, the zonules, and the hyaloid membrane. In his postmortem material, Daicker never observed the deposits beyond the pre-ora [1, pp. 257–258]. Usually they subside towards the posterior orbiculus.

A second and rare disorder is homocystinuria, leading to whitish degenerative precipitates on the ciliary epithelium of the pars plana.

Drusen of the Ora and Pars plana

In 20% of all eye globes, pearl-like spherical or polyspherical translucent structures are found along the ora, always beneath ora teeth or associated with their meridional extension anteriorly. Sometimes they are also found within pars plana ridges that originate from large ora spikes. Known as ora drusen or pars plana drusen, these concentrically arranged corpuscles are covered by a single layer of pigment epithelium. A pinch-off of these drusen into the space of Garnier or into the vitreous base at the ciliary body is also possible.

1 **Postmortem globe of a newborn:** Early cystoid degeneration of the peripheral retina in an otherwise fully differentiated globe.

2 Cystic ora as seen from above. Early cystoid degeneration, first involving the ora.

3 Cystic ora as seen from above with a 'spicule' on top; the cystoid degeneration extends towards the equator in a burrow-like fashion.

4 Cystoid retinal degeneration as seen from above: burrow-like spread in tongue-shaped areas.



- 5 View of advanced cystoid retinal degeneration (Blessig-Iwanoff cysts). Intraretinal cavities are crossed by pillars.

- 6 **Peripheral cystoid degeneration (Blessig-Iwanoff cysts):** View of the cystoid spaces. The retina below is detached by an artifact.

- 7 Isolated area of cystoid retinal degeneration surrounded by a retinal vessel peripherally. Note small retinal hole above.

- 8 In the inferior temporal quadrant there is a bleb of prominent cystoid degeneration 3 mm in height.



- 9 Polycyclic cobblestones (paving stones) within an area of cystoid retinal degeneration.

- 10 **Cystoid retinal degeneration with snail tracks and retinal rosettes (tufts):** Two large frayed retinal rosettes with vitreous condensations at the top are located between the equator and the ora. Behind the base of these two rosettes the retina reveals cystoid degeneration and yellow-whitish glistening spots of varying size in its inner layers.

- 11 Band-shaped tractional cystoid degeneration at the posterior border of the vitreous base.

- 12 **Cystoid degeneration caused by vitreoretinal traction:** There is a circular band of honeycomb formation in the retina between the equator and the ora at the posterior edge of the vitreous base.

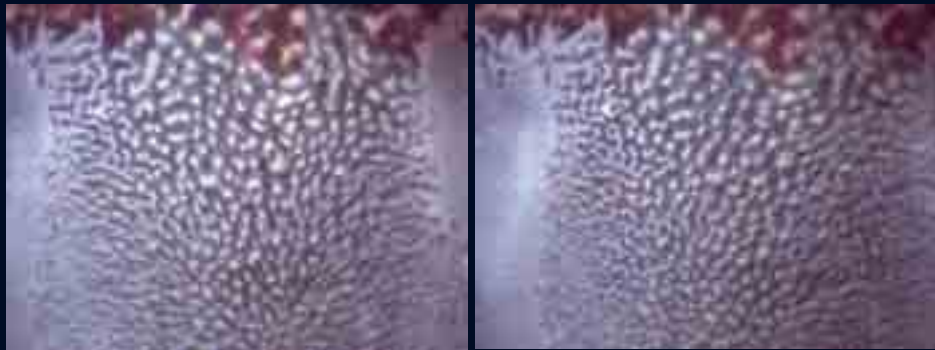


13 **Exposed cystoid retinal degeneration:** Vitreous traction at the posterior border of the vitreous base has opened up an elevated band of cystoid degeneration.

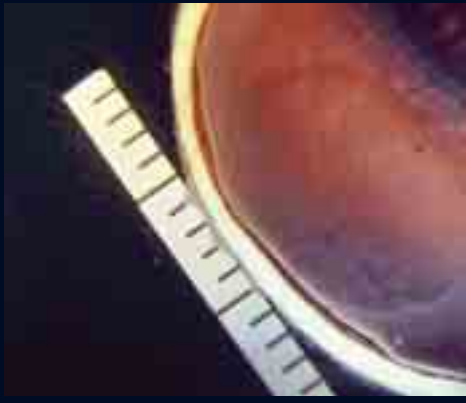
14 Between the temporal ora and equator there is an isolated cystoid, slightly prominent retinal lesion with perivascular holes of the inner retinal layers.

15 **Snail track with cystoid retinal degeneration:** Lobular cystoid degeneration behind the lower ora with disseminated fine glistening spots in the innermost and outermost retinal layer.

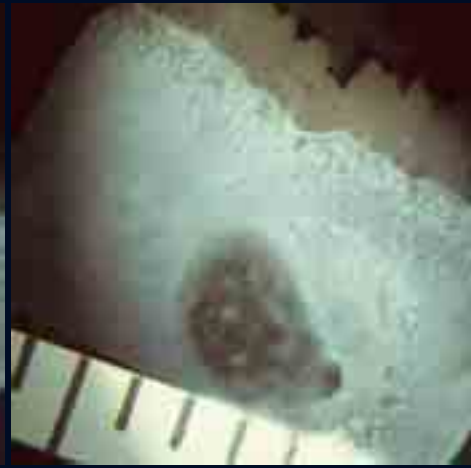
16 Marked peripheral cystoid retinal degeneration with adjacent pseudocystic degeneration of the pars plana.



- 17 Senile retinoschisis, forming a retinal cyst in an area of cystoid degeneration.
- 18 Seen from above, there is an almost round retinoschisis bleb within an area of cystoid degeneration, nearly reaching the ora (case 17).
- 19 In a transverse section (case 17) there is an impressive filiform elongation of the glial pillars with early retinal cleavage/schisis into two sheets.
- 20 An almost round retinoschisis bleb (as seen from above) 6.5 mm in diameter and 1 mm from the ora, next to an area of cystoid degeneration. A tiny retinal vessel can be seen on the inner leaf.



- 21 Senile retinoschisis of 3.5 mm diameter with extensive laminar loss of the outer leaf.
- 22 Large round bleb of retinoschisis as seen from above, with highly elongated filiform glial pillars still without complete separation of the two retinal sheets.
- 23 Multiple juxtavascular holes in an area of cystoid degeneration in a case of proliferative diabetic retinopathy.
- 24 **Snail track in retinoschisis:** There is a large bullous retinoschisis extending from the lower temporal equator up to the ora, the inner sheet and the inner part of its pillars being densely covered by snail track grains (transverse section).



25 **Snail track in retinoschisis:** Behind a massive cystoid retinal degeneration there is a retinoschisis 5 mm in diameter with a round hole in the outer leaf. The inner as well as the outer leaf contain many glistening spots.

26 **Congenital retinoschisis in a newborn with trisomy 13:** At the upper nasal periphery, close to the ora and extending to the equator, the retina is altered over a disc-like area 9 mm in diameter. Here, it is avascular, while along both margins the fetal vascular sprouts of the peripheral retina are visible. The margins of the disc are bulging into the vitreous. The base appears coarsely hammered.

27 Epithelium of the preretina with marked pseudocystoid degeneration, enclosing a small translucent pars plana cyst. Note the extensive cystoid degeneration of the retina behind the ora.

28 Flat whitish deposit of unknown etiology on the lower anterior pars plana in the space of Garnier.



29 On the lower pars plana, astride the ora, there are friable chalk-white masses.

30 Synchysis nivea particles can be found over the whole pars plana and pars plicata up to the band of Wieger.

31 Multiple almost confluent macrocysts, reaching the pars plicata anteriorly.

32 Multiple large pars plana cysts anterior to the temporal ora. Posterior to the ora, there is a band-shaped chorioretinal scar.



33 'Mushroom-shaped' pars plana cyst reaching from the ora to the pars plicata. Note the areas of cystoid degeneration.

34 On the temporal pars plana, there is a huge pars plana cyst with clear content, reaching the posterior end of the ciliary processes.

35 The pars plana is covered by huge confluent cysts with clear content, extending up to the posterior border of the pars plicata.

36 Large pars plana cyst with partially elevated pigment epithelium.



37 Anterior to the temporal ora, a few small pars plana cysts can be seen. Posterior to the ora, there is distinct cystoid retinal degeneration.

38 Temporal inferiorly there are multiple large pars plana cysts with a clearly visible midline.

39 Pars plana cysts anterior to a scarred pre-ora temporally below.

40 Opaque cysts of the nasal upper pars plana, not quite passing over the mid pars plana.



41 Multiple clustered vesicular and partially indented pars plana cysts with whitish content.

42 Small to medium-size cobblestones with discrete foci of coarse hyperpigmentation directly adjacent to the ora.

43 Coalescing polycyclic cobblestones behind the ora with several large choroidal vessels visible in between.

44 Large area of cobblestone degeneration with individual polycyclic foci coalescing and numerous small nodular drusen in between. Linear vascular degeneration between the cobblestone degeneration and the ora.



45 At the temporal ora there are large polycyclic aggregations of cobblestone degeneration with two small pars plana cysts at the anterior border.

46 Cobblestone degeneration within an area of peripheral cystoid degeneration. Directly above the large cobblestone is a small retinoschisis.

47 **Etat craquelé of the peripheral pigment epithelium (same case as in fig. 46):** The retina with cystoid degeneration has been folded backwards to the left towards the ora. Next to the large cobblestone degeneration there is a linear degeneration with a corresponding retinal degeneration within the cystoid area.

48 **Linear degeneration of the retinal pigment epithelium:** Etat craquelé: The peripheral pigment epithelium appears mottled and permeated by pale lines as if it was disrupted.

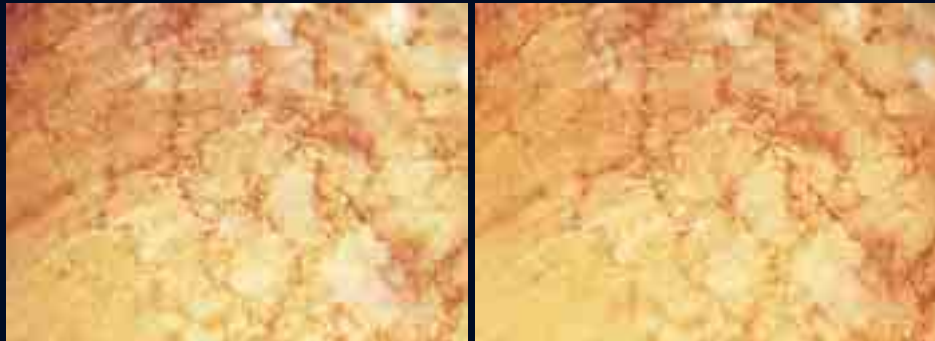
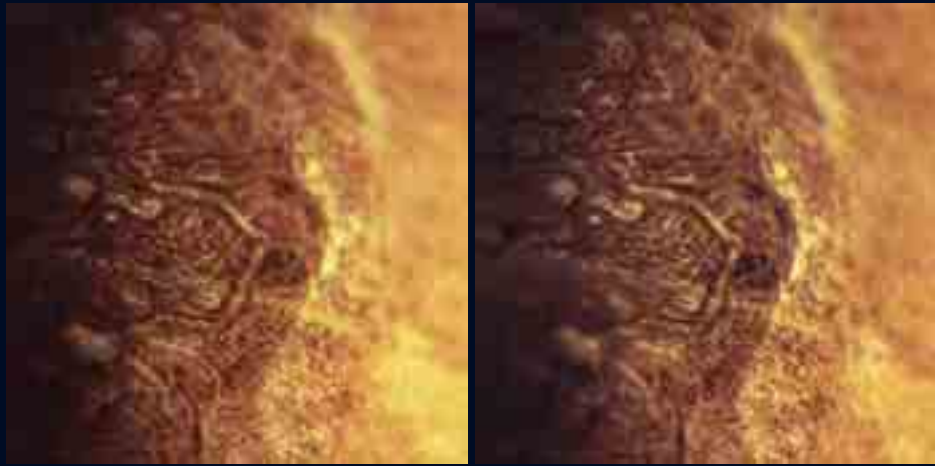


- 49 Aspects of a linear vascular degeneration between état craquelé and the ora as seen from above.

- 50 **Linear vascular pigment epithelial degeneration:** The peripheral retina shows an impressive circular état craquelé. Temporal superiorly there is a bulging subretinal vessel running along the extension of a prominent radial pars plana vessel meridionally as far as the equator where it branches out. The overlying retinal pigment epithelium is linearly mottled and degenerate.

- 51 **Reticular pigment epithelial degeneration:** At the equator, there are hyperpigmented lines forming a polygonal network. In between the cells, along the lines, nodular hyaline drusen are seen.

- 52 Extensive cystoid degeneration of the peripheral retina with a relatively well-demarcated retinal excavation.

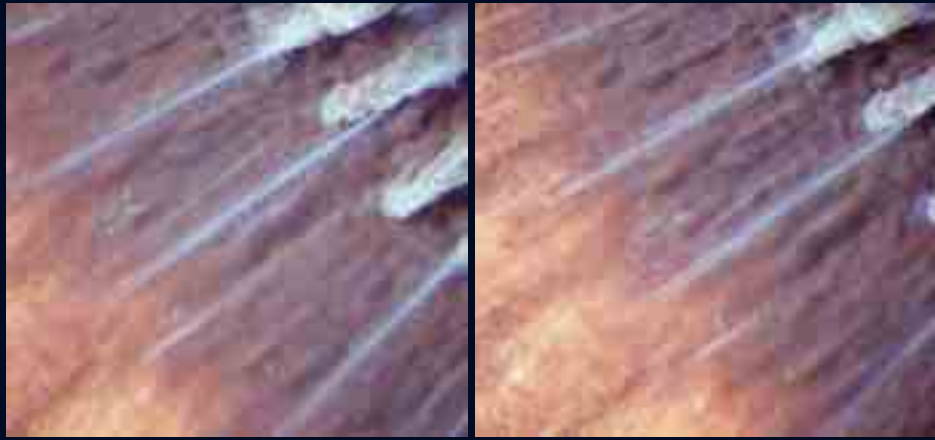


53 **Pseudoexfoliation:** There are massive deposits of Busacca covering the zonules in the space of Garnier. They appear as a sort of 'dusting' of the fine filaments.

54 **Pseudoexfoliation:** Extensive deposits of Busacca cover the surface of the ciliary body and the zonules as well as the posterior iris surface.

55 **Pseudoexfoliation:** At the insertion into the lens capsule the zonules are densely covered by Busacca deposits. They can easily be traced back along their course between the ciliary valleys.

56 Discrete pars plana druse within a pars plana ridge (2-year-old child). Adjacent more pars plana ridges and a lissora can be seen.

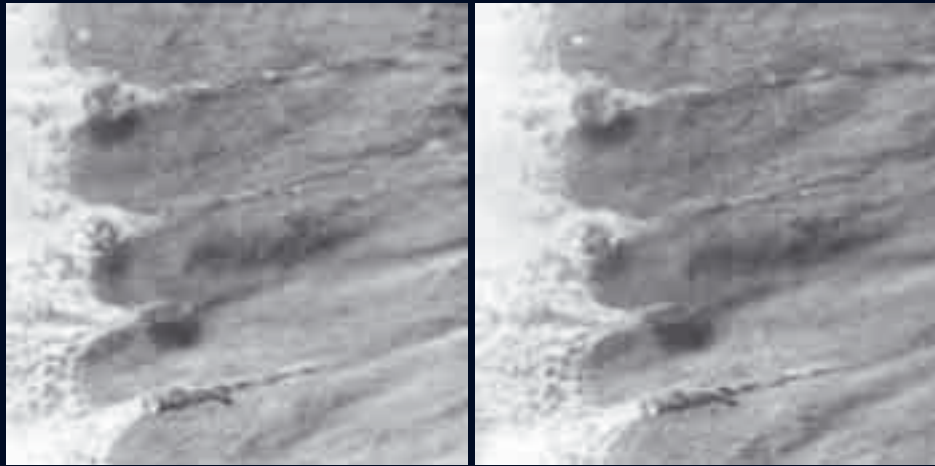


57 Two drusen located within ora teeth, the upper one polyspherical, and both partially covered by pigment. There is a hint of pars plana ridge formation.

58 A small druse in a crista ciliaris. In addition, an état craquelé can be seen circumferentially.

59 **Peripheral fundus with drusen:** Etat craquelé of the pigment epithelium behind the ora with multiple nodular drusen of varying size.

60 Disseminated nodular drusen of the peripheral choroid, overlying an area of large confluent cobblestone degeneration with linear pigment irregularities.





Inflammatory Lesions

Introduction

Uveitis is the most common inflammatory lesion of the peripheral fundus. Pure retinitis or vitreoretinitis limited to the peripheral fundus is comparatively rare.

During inflammation, it is difficult to view the fundus despite modern devices and improved examination techniques. The most important limiting factor relates to clouding of the media associated with the inflammatory process, especially vitreous opacities. For the clinician, peripheral vitreous condensations indirectly point towards an inflammation of the underlying pars plana.

For a detailed histopathological examination of such an inflamed pars plana, the infiltrated peripheral vitreous must be separated and removed, thereby giving rise to even more (and possibly artifactual) destruction of the inflamed region.

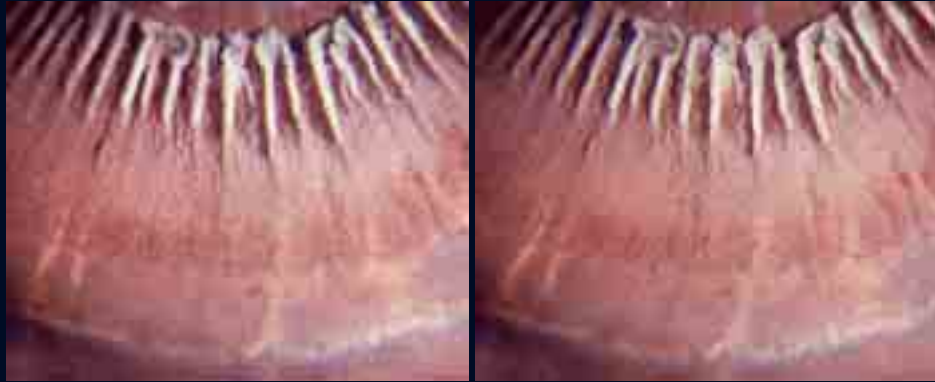
Thus, Daicker's material provides only few pictures with acute exudative inflammation [1, pp. 166–171]. In most cases, late changes such as scars and fibrous metaplasia are illustrated. A topographical localization of the inflammation can be defined as follows: cyclitis posterior: corresponds to orbiculitis; most peripheral chorioretinitis: localization behind the ora; and orbiculo-chorioretinitis: localization along the ora [1, p. 170].

Severe inflammation of the orbiculus (pars plana) with alterations in both pigmented and non-pigmented ciliary epithelium is followed by atrophic scar formation. The destroyed epithelium is replaced by fibrovascular scar tissue, frequently infiltrated by pigmented cells, and that tissue often reveals a mild lymphocytic infiltrate even long after the acute event.

The scar is located on the anterior orbiculus, thus involving the zonular architecture (fig. 12). In case of a preretinal scar, the vitreous is firmly attached.

Persisting severe inflammation can lead to tractional retinal detachment due to organization of the exudates and scar contraction in the orbicular space and the anterior vitreous, which eventually form a cyclitic and retrolental scar membrane. This finally leads to a phthisical eye.

- 1 **Iritis due to streptococcal sepsis:** Posterior chamber with whitish-yellow precipitates.
- 2 **Sequelae after iritis:** Pigment dispersion into the lower space of Garnier. Hyperplasia of ciliary epithelium.
- 3 **Septic chorioretinitis in a case of pneumococcal meningitis:** The vitreous is cloudy with cellular infiltrates. Small foci of exudate and pus are overlying the pars plana, indicating a focal posterior cyclitis. The lower pars plana and the valleys between the ciliary processes are covered by a cellular exudate.
- 4 **Granulomatous retinitis (sarcoidosis):** The pars plana is covered by epiciliary precipitates, the peripheral retina and the ora serrata by a fibrinous membrane.



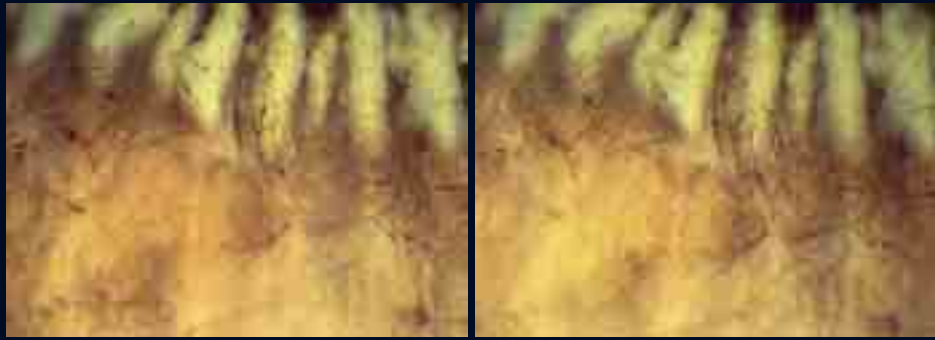
- 5 **Mild chronic cyclitis:** There are whitish crumbly masses lying like a snow bank in the vitreous base on the inferior pars plana.
- 6 **Cell precipitates and exudates in the space of Garnier in a case of panophthalmitis:** The anterior vitreous is cloudy due to dot-like and diffuse opacification. Behind the lower edge of the vitreous base there are precipitates on the inner retinal surface.
- 7 **Severe endophthalmitis:** Anterior and posterior chamber are filled with white-yellow exudate. The vitreous shows a synchysis nivea. Along the ora serrata, a band of cobblestone degeneration is seen.
- 8 **Fungal endophthalmitis:** There is a white fluffy ball of fungal elements (about 3 mm in diameter) on the pars plana.



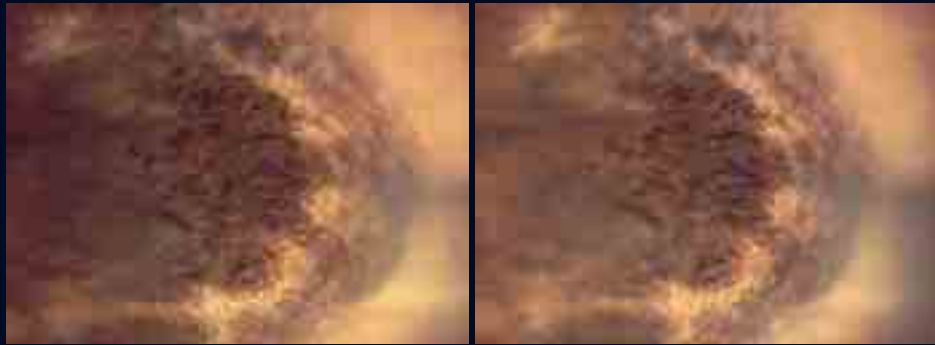
- 9 **Pars plicata:** Whitish-felty cyclitic membrane with corresponding circumscribed cataract of the posterior lens cortex.
- 10 **Spontaneous phthisis: 'Callous floater':** Above the ciliary processes and the posterior iris surface there is a whitish solid nodule (1.5×2 mm).
- 11 **Pars plana/pars plicata:** Whitish membranes surrounded by fine strands of proliferating ciliary and pigment epithelium.
- 12 **Pars plana/pars plicata/lens:** Extensive fibrous membranes with strand-like proliferation of ciliary and pigment epithelium. Cataracta complicata.



- 13 **Lower and nasal pars plana after cyclitis:** Fibroepithelial scars with bone spicule-like pigmentation.
- 14 **Inferotemporal pars plana:** Depigmented scars due to herpes zoster ophthalmicus.
- 15 **Findings after peripheral chorioretinitis and peracute posterior cyclitis:** There are irregularly shaped whitish spots on the lower orbiculus, 1–2 mm in diameter, that are partially lined by pigmentary disturbance; some appear slightly elevated with button-like proliferation of the ciliary epithelium.
- 16 Fibrous cicatrization overlying the pars plana with retinal traction.



- 17 **Pars plana scars and cysts:** Dot-like fibrotic scars of the temporal pars plana. Large pars plana cysts along the whole temporal circumference.
- 18 **Findings after peripheral chorioretinitis and iridocyclitis:** The whole upper retinal periphery reveals broad geographic white granular chorioretinal scars, partly extending onto the pars plana. Some individual pars plana cysts can be seen.
- 19 **Posterior ora:** Fibrous tissue and pigment epithelial proliferation forming a 'Ringschwiele'.
- 20 **Ora and posterior pars plana:** Scar with pigmentary disturbance and strand-like proliferation of the pigment epithelium.

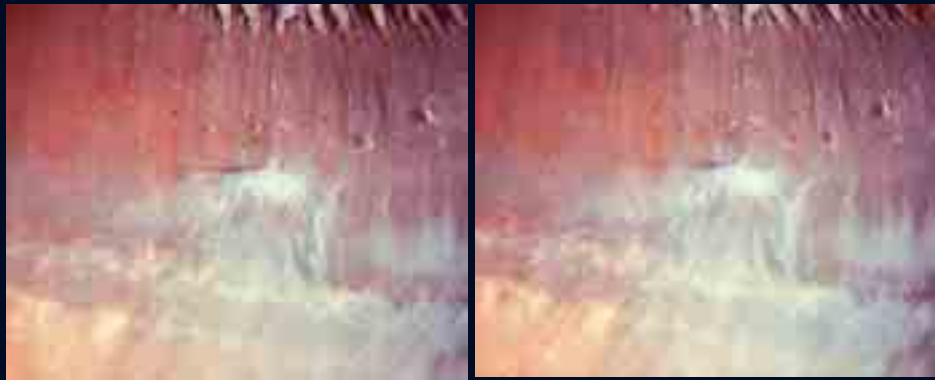


21 Preretinal scar with nub-like hyperplasia of the ciliary epithelium and cystoid degeneration of the peripheral retina.

22 **Clinical iridocyclitis with secondary cataract:** There is a feathery scar on the posterior pars plana at the 6 o'clock position with traction on the peripheral retina.

23 **Depigmentation after chorioretinitis and cyclitis:** On the nasal side, from the pars plana to the equator, there is a large atrophic whitish band-shaped chorioretinal scar (pseudocoloboma) with heavily pigmented edges.

24 **Upper temporal ora serrata:** Chorioretinal scars caused by clinically diagnosed herpes zoster ophthalmicus.





Tumors

Introduction

Hyperplasia of the Nonpigmented and Pigmented Pars plana Epithelium

Extensive hyperplastic changes are seen more frequently in the nonpigmented than the pigmented ciliary epithelium of the orbiculus. These are seen as individual or collections of epithelial buds on the inner surface of the ciliary epithelium. Histologically, these buds are composed of epithelial cells with indistinct cell borders.

Reactive focal pseudoadenomatous hyperplasia of the pars plicata (Fuchs adenoma) can be found in 25% of all people as nodular round whitish-gray tumors (1 mm in diameter) of the pars plicata.

The pigment epithelium can proliferate in an endophytical fashion in multiple layers towards the inner globe, thus creating a pigmented cushion beneath the regular or altered ciliary epithelium.

Excrescences of the pigment and ciliary epithelium on the orbiculus seen in a posttraumatic 'Ringschwiele', after inflammation or after long-standing retinal detachment, will be discussed in the respective chapters.

Tumors of the Most Peripheral Fundus (Fundus extremus)

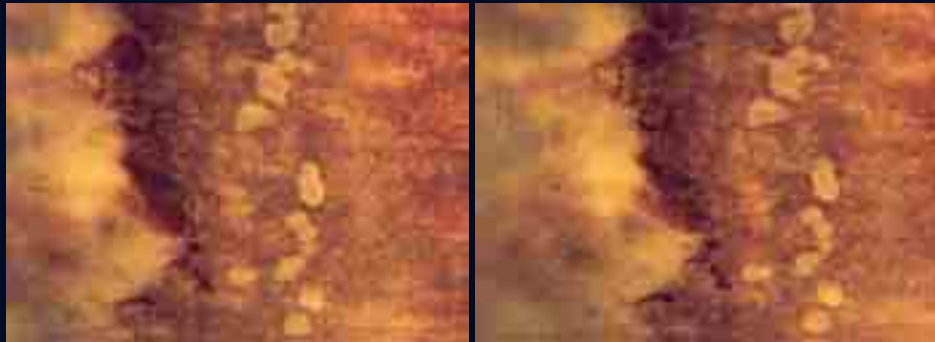
The most frequent malignant tumor of the uvea, the malignant melanoma, is also the most frequent tumor type of the fundus extremus. Daicker in his postmortem material found about 7% of small uveal melanomas arising from the posterior ciliary body [1, p. 274]. Because of their peripheral location, they are detected and treated not

until late in the course of the disease. Structure and histology are identical to those of uveal melanomas. The configuration of melanomas in the posterior ciliary body tends to be nodular rather than flat. As a consequence, a spherical swell protrudes into the vitreous and against the lens with subsequent subluxation. In about half of the cases, the overlying ciliary epithelium shows a shallow serous or hemorrhagic detachment.

Tumors of the retina like the often multicenter retinoblastoma or neuroepithelioma can also exhibit segregated foci at the ora. Even metastases from such tumors can be found at the ora via extension along the subretinal space. Hematogenous metastases at the orbiculus are infrequent compared with those in the posterior uvea. As typical examples, metastatic tumors of the fundus extremus arising from a carcinoma of the thyroid (fig. 26), of the breast (fig. 28), and of the bronchus (fig. 27) are illustrated.

Tumors arising from the stroma of the orbiculus like leiomyoma, hemangioma, neurofibromatosis, and xanthogranuloma are rare. Tumors of the hemopoietic system like leukemia, oculocerebral non-Hodgkin lymphoma or plasmocytoma can give rise to an infiltration of or a hemorrhage into the space of Garnier as well as to deposits on the pars plana.

- 1 Marked button-like hyperplasia of the ciliary epithelium of the anterior pars plana.
- 2 The pre-retina shows multiple button-like excrescences of the ciliary epithelium and, immediately adjacent to the ora, also of the pigment epithelium.
- 3 Button-like excrescences of the ciliary epithelium, occasionally projecting through the zonules, and a proliferation of pigment epithelium next to the ora (detail of fig. 2).
- 4 Anterior to the temporal ora there are nodular proliferations of pigment epithelium in the pre-retina. Note extensive cystoid retinal degeneration.



5 Small nodules of ciliary epithelium extend anteriorly along the pars plana ridges (same case as in fig. 4, detail).

6 A pigment epithelial morula in the space of Garnier below. Short ciliary processes of the third order on the anterior orbiculus.

7 Freely floating morula of the pigment epithelium above a third-order ciliary process. Further anteriorly in between the ciliary processes there is a Fuchs adenoma.

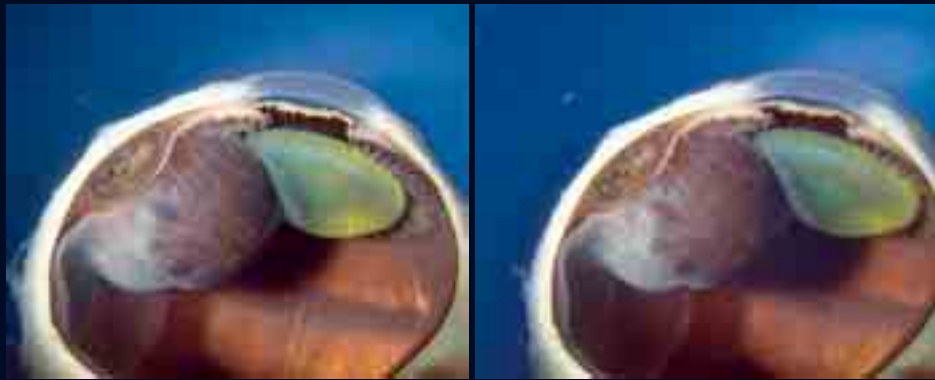
8 Small spherical Fuchs adenoma of the ciliary body.



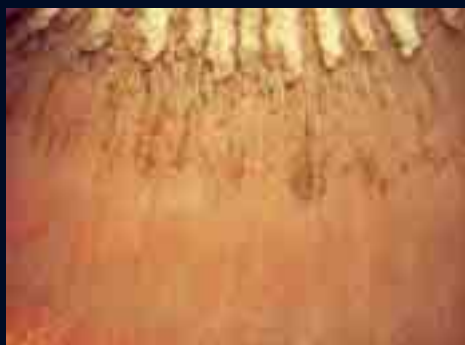
- 9 Fuchs adenoma of the ciliary body lying between two ciliary processes.
- 10 Fuchs adenoma of the ciliary body, located at a superonasal ciliary process.
- 11 Two morula-like whitish excrescences at the temporal ciliary processes (Fuchs adenomas), reaching the posterior iris surface.
- 12 Complete iris coloboma and aphakia. In the area of the coloboma above there is a large hyaline ciliary nodule with disseminated chalky spots (ciliary adenoma?).



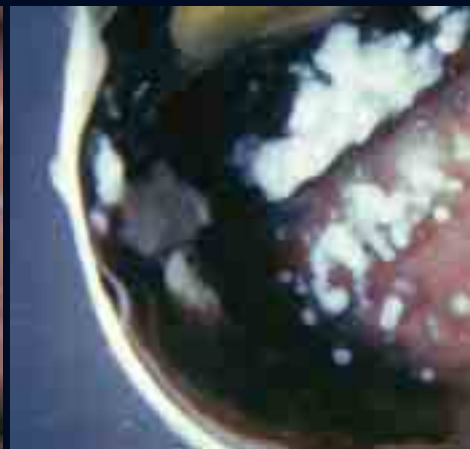
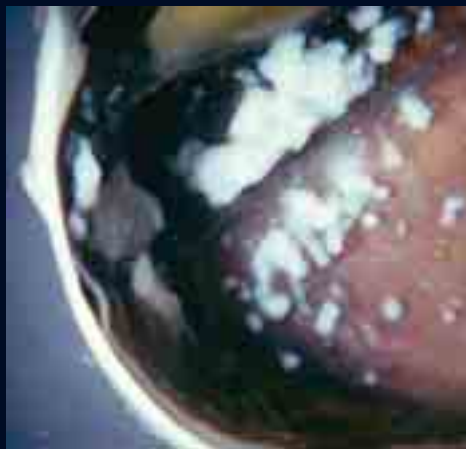
- 13 Cysts of the ciliary epithelium with clear content, located in between the ciliary villi in the pars plicata.
- 14 In the lower temporal ciliary body there are two ciliary epithelial cysts and a small Fuchs adenoma.
- 15 **Ciliary body melanoma:** In the lower ciliary body there is a large brown nodular tumor approximately 9 mm in height. Its apex indents the lower lens equator and pushes the lens slightly upwards. Posteriorly, the tumor reaches the equator. Behind it, the retina shows a narrow crescent-shaped collateral serous detachment.
- 16 **Ciliary body melanoma:** The cut surface appears partly solid, partly composed of lumps of brownish cells surrounding optically empty spaces or spaces filled with serous fluid (same case as illustrated in fig. 15).



- 17 **Ciliary body melanoma:** In the temporal ciliary body and anterior uvea there is a nodular tumor, measuring about 16 mm along the equator and 10 mm in height.
- 18 **Ciliary body melanoma:** The temporal anterior part of the globe reveals a bullous detachment of the ciliary epithelium (right-hand side) measuring 5×5 mm. The temporal ciliary body in its folded part is elevated over an area measuring also 5×5 mm, with a light brown cauliflower-like tumor of 3×5 mm arising from there. The equator of the lens is indented by the tumor apex. Posteriorly, the tumor reaches the pars plana where it is visible at the bottom of the ciliary epithelial detachment.
- 19 After removal of the lens the tumor can be seen extending onto the iris for 1.5 mm. The iris pigment epithelium in this area shows circular folds while these are otherwise meridional (same case as in fig. 18).
- 20 Ciliary body melanoma with pigment seeding into the space of Garnier below (same case as in fig. 18).



- 21 **Ciliary body melanoma after treatment with proton beam irradiation 14 months previously:** Within the nasal ciliary body, extending back to the equator beneath the retina, there is a nodular brown tumor 9.5 mm in diameter and approximately 7 mm in height. The tumor surface is speckled whitish.
- 22 **Melanocytoma of the ciliary body:** Piece of corneoscleral tissue 6 mm in diameter. On the inner aspect the pars plicata of the ciliary body is spheroidally elevated.
- 23 **Melanocytoma of the ciliary body:** Arising from within the center of the ciliary body there is a black nodular tumor measuring 3 mm meridionally and 2 mm radially. Anteriorly, it reaches the chamber angle and extends into the iris root as well as into the sclera at the site of the trabecular meshwork (same case as in fig. 22).
- 24 **Peripheral retinoblastoma with massive seeding into the anterior vitreous over the pars plana:** There are masses of large white balls and sheets of disseminated tumor cell nodules on the lower peripheral retina, especially on the lower pars plana. They measure 0.5–3 mm in diameter.

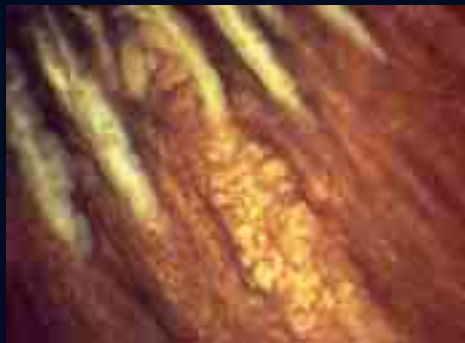


25 **Retinoblastoma:** After opening of the globe a total retinal detachment can be seen. On the inner as well as particularly on the outer surface of the retina extensive nodular whitish tumor masses are present, interspersed with porcelain-white granules. In addition, tumor seedings are also seen adjacent to the upper zonules and along the posterior lens surface.

26 **Metastasis of a medullary thyroid carcinoma in the pars plana:** On the pars plana there are disseminated streaks of whitish subepithelial nodules.

27 **Ciliary body metastasis of a bronchus carcinoma:** Whitish fluffy tumor excrescences between the ciliary processes and over the pars plana. Flat cystic elevation of the ciliary epithelium on the respective pars plana.

28 **Choroidal/ciliary body metastasis of a breast carcinoma:** The lower and lateral ciliary body is enlarged by tumor masses from the choroid, and a lens-shaped choroidal tumor is seen at the temporal equator. The inner surface of the ciliary body and pars plana appears almost normal.



29 **Ciliary body metastasis of an undifferentiated carcinoma:** The lower globe reveals a nodular tumor 16 mm in diameter and 11 mm in height beneath the ora, extending from the pars plicata until behind the equator. In addition, there is a collateral bullous retinal detachment.

30 **Retinal lymphoma after irradiation:** There are whitish ant-like pearls on the lower pars plana.

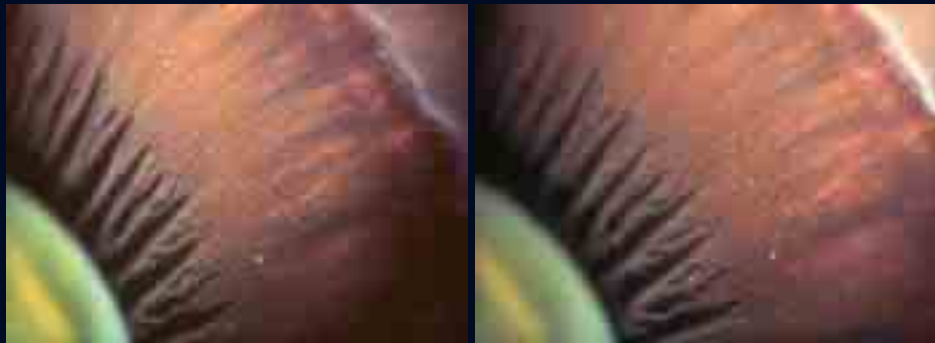
31 Single small pars plana cysts on the middle pars plana without contact to the ora in a case of plasmocytoma.

32 Multiple opaque pars plana cysts next to the ora and further anteriorly in a case of plasmocytoma (same case as in fig. 31, contralateral eye).



- 33 **Leukemic hemorrhage:** Between the lower ora serrata and the equator there is a multinodular intraretinal hemorrhage with whitish dots and a bleeding into the vitreous base at this site.

- 34 **Deposits in acute myeloic leukemia:** On the lower pars plana in the space of Garnier, between the zonules, and in the valleys between the ciliary processes there is a dissemination of yellowish glistening dots the size of cellular aggregations.





Traumatic Lesions

Introduction

Contusions deform the ocular globe depending on the force involved as well as on the velocity. In the region of the fundus extremus, contusion damage usually occurs at the insertion of the ciliary body (at the scleral spur), at the suspension of the lens, at the 'insertion' of the retina at the ora serrata, and at the insertion of the anterior and posterior hyaloid membranes [2, p. 75].

The identification of traumatic alterations in the region of the fundus extremus is important since blunt trauma can lead to serious complications such as retinal detachment, proliferative scar formation, and secondary glaucoma.

Posttraumatic Peripheral Vitreous Detachment

After blunt trauma, small vitreous detachments occur quite frequently, as is obvious from pre-retinal hemorrhage. A large rhegmatogenous vitreous detachment can also be caused by ocular contusion. Degenerative factors in the vitreous cortex due to age or myopia play an additional role. A traumatic etiology can thus be postulated only on the basis of a temporal correlation [2, p. 75].

Tear Formation of the Fundus extremus

Immediate traumatic tears can be found in the most peripheral retina, at the ora (ora tear) and in the ciliary epithelium. Indirect traumatic defects originate from tractional scars or from preexisting alterations of the most peripheral retina and vitreous. About 12% of all retinal tears are located at

the ora [9]. Such tears are seen mostly in children and young adults. The tears form predominantly at the anterior or posterior border of the vitreous base. They run more or less parallel to the ora and are simple or arborescent [2, pp. 76--78]. They are preferentially found in the temporal periphery. Eventually, posttraumatic scar contraction in the anterior globe or organized vitreous condensations can lead to new tears or an extension of pre-existing tears even after months or years. Severe contusion can also cause disruption of the pigment epithelium particularly in the area of the orbiculus, which is frequently associated with massive hemorrhage into the orbicular space or into the vitreous. Perforating injuries can lead to disruptions at any site of the peripheral retina, the orbicular ciliary epithelium or the pigment epithelium.

Traumatic Detachment of the Inner Ocular Layers of the Fundus extremus

A posttraumatic detachment is seen primarily after tearing of the retina and secondarily after proliferative changes. In 15% an additional detachment of the ciliary epithelium with smooth transitions can be found [10]. In those cases the vitreous base plays an important role as a scaffolding. After vitreous prolapse or shrinkage towards the front, the entire vitreous base, i.e. the vitreous along with an adherent strip of tissue from the ciliary epithelium and the retina, is pulled off. Thus, at the same time, approximately parallel tears form due to traction at the anterior and posterior border of the vitreous base.

Traumatic Detachment of the Pars plana

Direct or contrecoup trauma, perforating injuries of the lateral globe and cyclodialysis can lead to a total detachment of all tissues of the pars plana corporis ciliaris. Similar to a choroidal detachment elsewhere, there is a serous-hemorrhagic separation of the subchoroidal fibrous lamellae in addition to the outer layers of the ciliary muscle. A smooth separation between the sclera and the ciliary body is comparatively rare.

Clinically a surprisingly fast and complete resorption of such traumatic choroidociliary detachments can often be observed. Larger lesions, after organization, can lead to callous scars.

Proliferative Posttraumatic Reaction (Ringschwielen Formation)

When there are defects in the retinal pigment epithelium near the ora or at the orbiculus, occasionally a reactive and reparative infiltration of inflammatory cells, macrophages, fibroblasts, and vascular sprouts from the uveal stroma can be observed. This tissue grows beneath the inner sheet of the ocular cup and, in case of breaks of the latter, also towards the inner part of the eye. Here there will be organized hemorrhage.

Scar formation by proliferation of the pigment epithelium and/or the nonpigmented ciliary epithelium is frequently seen after trauma in the area of the ora and the orbiculus.

Neovascularization of the vitreous base is seen after long-standing retinal detachment reaching up to the ora. Originating in the uvea, new vessels develop between the detached retina and the

epithelium of the pre-retina. At the ora, Daicker observed in older individuals physiological choroidal vessels within the elastic lamina just beneath the pigment epithelium, allowing easy access of vessels into the vitreous [1, p. 159].

The so-called Ringschwielen formation beneath the retina can be caused by traction of the completely detached retina, and possibly the commonly found preexisting physiological degenerative changes of the pigment epithelium in this area are relevant.

Retinal detachments with complete elevation of the orbicular epithelium bring about cushion-like scars that extend from the ora posteriorly beneath the anterior edge of the ciliary detachment. These scars are arranged in a circular fashion along the posterior orbiculus and are therefore called Ringschwiele.

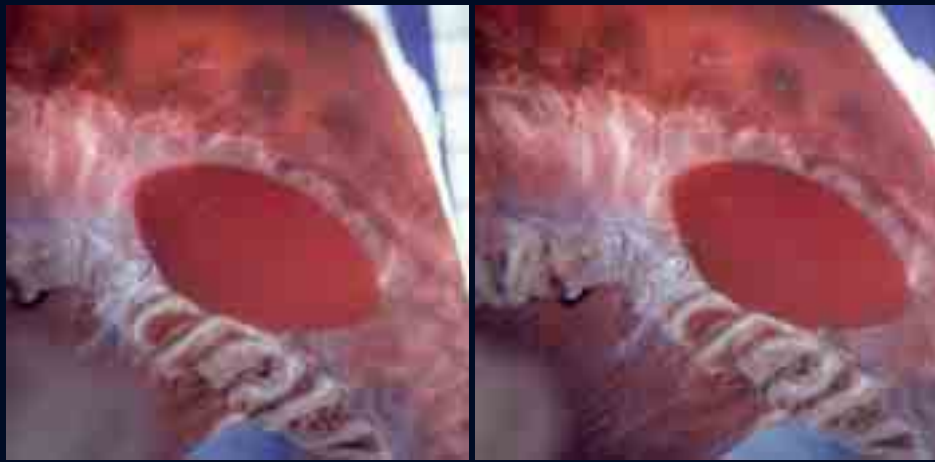
After injury to the pre-retina with trauma to the nonpigmented and pigmented ciliary epithelium scars can develop in the space of Garnier with proliferation of the pigmented and nonpigmented epithelium together with fibrous tissue and vessels forming a jagged cushion-like scar as described above.

1 Ora tear with hemorrhage into the space of Garnier.

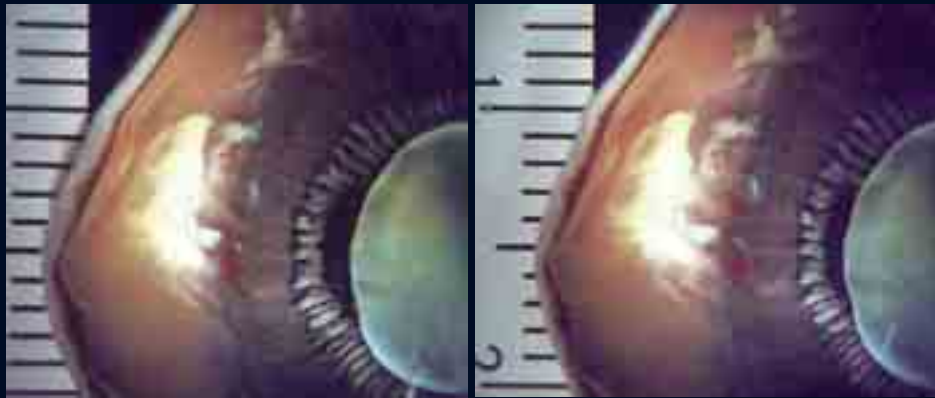
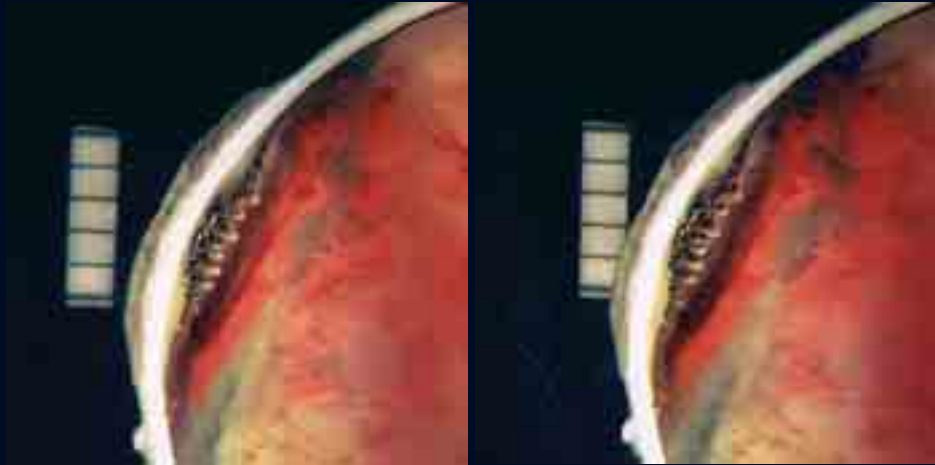
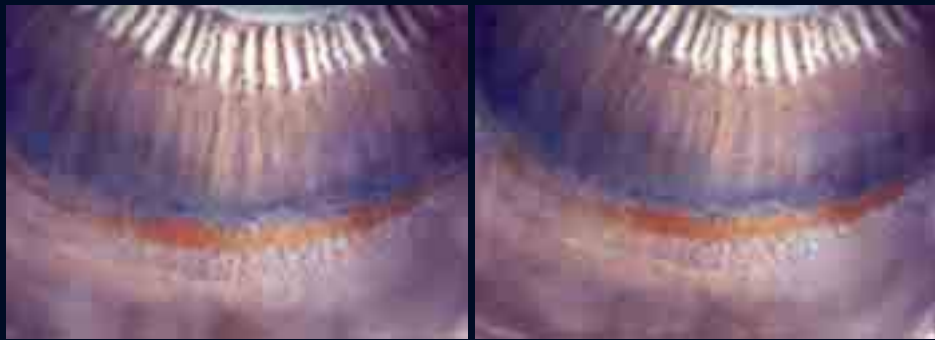
2 **Old ora tear:** On the temporal side, there is an ora tear of 1.5 mm in length with slightly elevated edges. Anterior to the tear, there are proliferations of the ciliary epithelium on the pars plana.

3 Traumatic retinal tear at the vitreous base.
Adjacent extensive cystoid retinal degeneration.

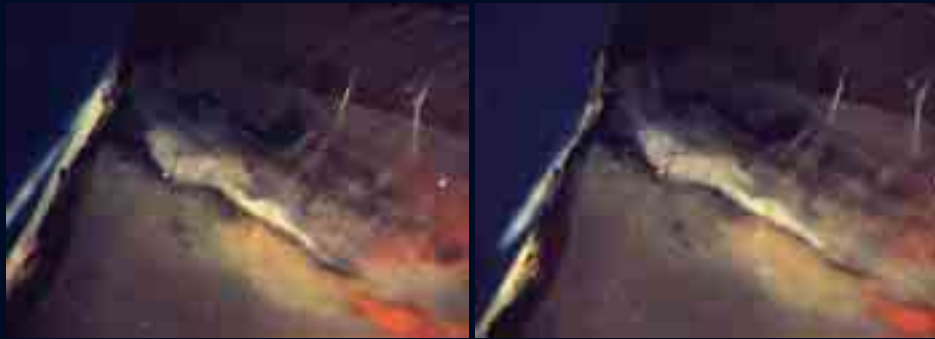
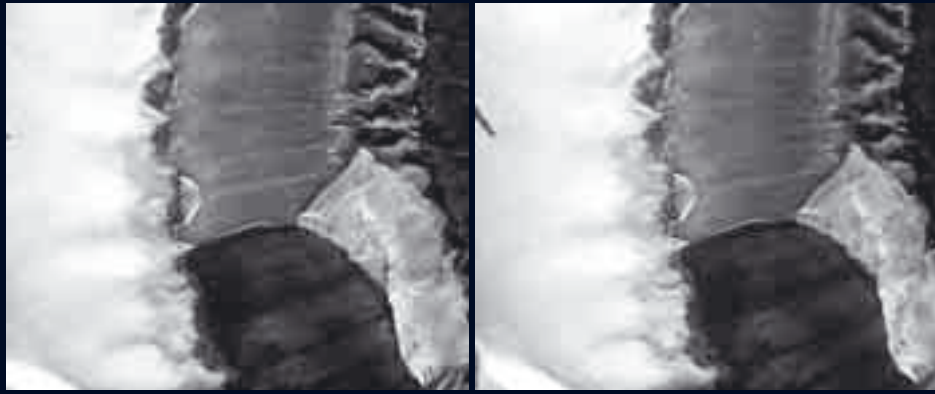
4 Pars plana hemorrhage 14 days after perforating injury.



- 5 A widespread sickle-shaped accumulation of liquid blood in the vitreoretinal recess at the level of the ora. Next to it, extensive cystoid retinal degeneration is present.
- 6 A dense recent hemorrhage at the vitreous base.
- 7 **Damaged zonules and iris prolapse into the posterior chamber after recurrent vitreous hemorrhage:** Partly organized next to recent hemorrhage in the vitreous base. Defective zonules over 4 mm with posteriorly translocated ciliary processes, flattening of the lens equator, and posterior iris prolapse.
- 8 Retina detachment at the ora or pre-ora. Areas with spots of pigment epithelial disruption beneath the ora and the pre-retina with exposure of the choroid.



- 9 **Status after severe blunt trauma (9-year-old child):** Recent wide-stretched tear of the ciliary epithelium anterior to the ora extending up to the pars plicata with rolling in of the ripped-off epithelium. There is no visible lesion of the pigment epithelium.
- 10 Disruption of the pre-retinal epithelium and parts of the pigment epithelium. Some areas of bare choroid are seen. The retina is torn off at the ora.
- 11 **Trauma due to windshield injury:** On the nasal side there is an incomplete partially calcified annular cataract. The ciliary processes have prolapsed anteriorly.
- 12 **Status after old corneal perforating injury:** Calcified traumatic cataract. The retina is drawn across the pars plana towards the lens by intra-ocular scar tissue. Adjacent, there are strands and nodules of proliferating ciliary epithelium.



- 13 **Epiciliary callous scar after perforating injury about 20 years previously:** There is aphakia with an extensive callous scar in the upper posterior chamber as well as behind the pupil.

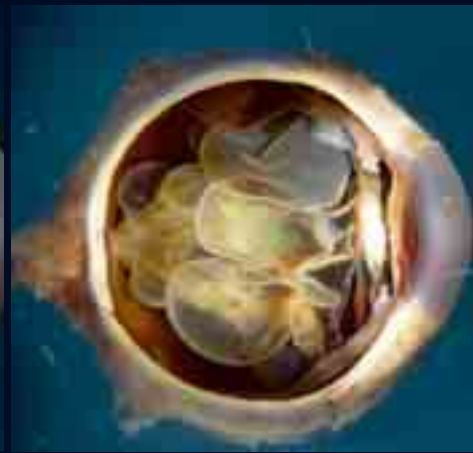
- 14 Meridional folds of the pars plana ciliary epithelium after retinal detachment. Adjacent, there are two pars plana cysts and a nodular proliferation of the pre-retinal epithelium.

- 15 **Luxation of the lens in early phthisis:** The globe reveals a distinct corectopia and a markedly constricted pupil. Below, the ciliary processes are pulled slightly towards the posterior iris surface, and the pre-retinal epithelium shows extensive nodular proliferations.

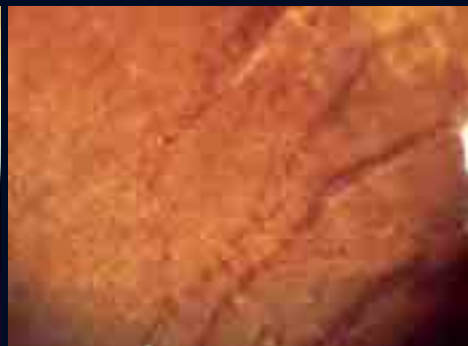
- 16 **Phthisical eye after long-standing total retinal detachment with advanced retinal atrophy:** In the peripheral retina, giant drusen can be seen on the choroid.



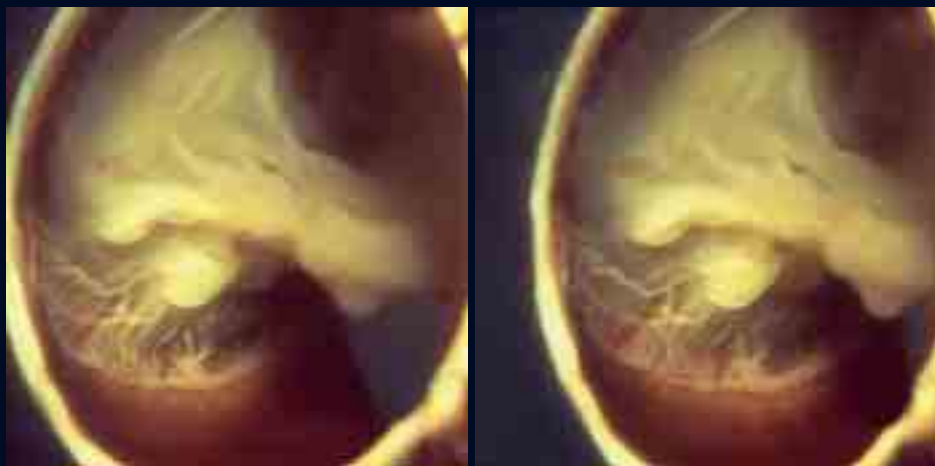
- 17 **Detail of figure 16:** Accumulation of variably sized drusen on the choroid, some partially pigmented, the larger ones free from pigment.
- 18 **Macrocystic retinal degeneration:** There is a complete retinal detachment with grape-like macrocysts.
- 19 **Etat craquelé after total retinal detachment:** Next to the detached retina there is a band-shaped area of linear degeneration.



- 20 **Pigmentary changes after blunt trauma:** On the pars plana the pigment epithelium and ciliary epithelium have proliferated in streaks. At the ora, several blot-shaped pigment irregularities are found.
- 21 **Pigmented chorioretinal scar:** Broad fleam-shaped black blotchy scar, 4 × 9 mm in diameter. There is a hyperpigmentation of the subretinal tissue and a pseudopigmentosa-like pigment migration, mostly perivascular, into the retina.
- 22 **Linear degeneration of the peripheral pigment epithelium after choroidal detachment:** Hyperpigmented parallel lines, merging near the site of the vortex veins.
- 23 **Perforating scar of the pars plana:** On the temporal pars plana, there is an atrophic choroidal scar 1 mm in diameter. Cataracta corticalis anterior et posterior.



- 24 **Status after perforating injury:** Perforating corneal scar. Slit-like defect of the iris pigment epithelium. Depigmentation of the ciliary processes with fan-like fibrous strands in the corresponding sector. The lens reveals a crescent-like indentation at the equator.
- 25 **Detail of figure 24:** Fan-like fibrous strands originating from the pars plicata at this site extend across the pars plana to an atrophic scar behind the ora.
- 26 **Status after corneal perforation:** A fibrous callous scar between the pars plicata and the ora at the 5 o'clock position has led to a pointed retinal traction. Adjacent, there is an incomplete ring-shaped secondary cataract with the most prominent lens remnants being present nasally.
- 27 **Status after perforating limbal injury:** There is a complete funnel-shaped retinal detachment including the ciliary epithelium of the pars plana with tractional tears of the detached ciliary epithelium.

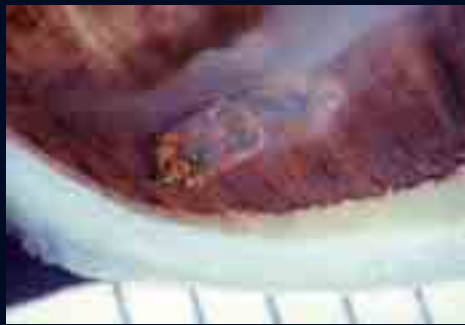


28 **Status after surgery for perforating injury:**
Funnel-shaped retinal detachment; the lower retina is more highly elevated, lying over the horizontal meridian and being drawn into the perforation site. The ciliary epithelium is also detached and has together with the retina prolapsed inversely over the ciliary body towards the cornea.

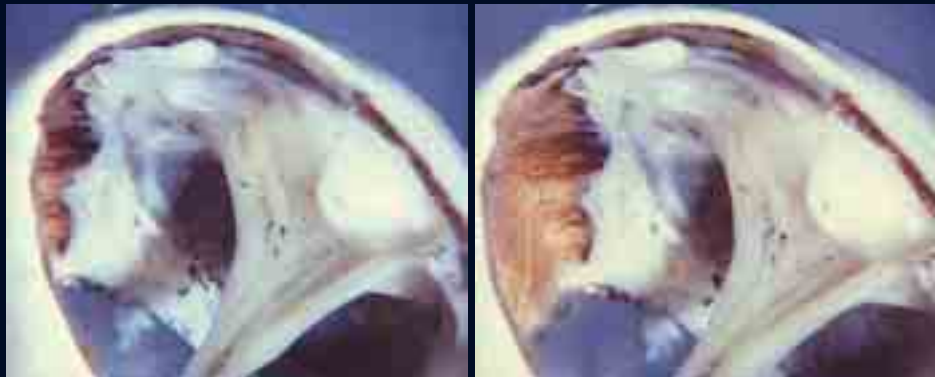
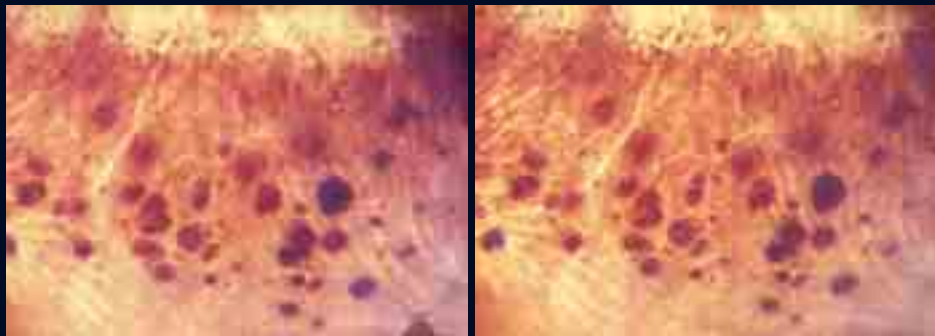
29 **Status after perforating injury:** Variably pronounced humps of pigment epithelium.

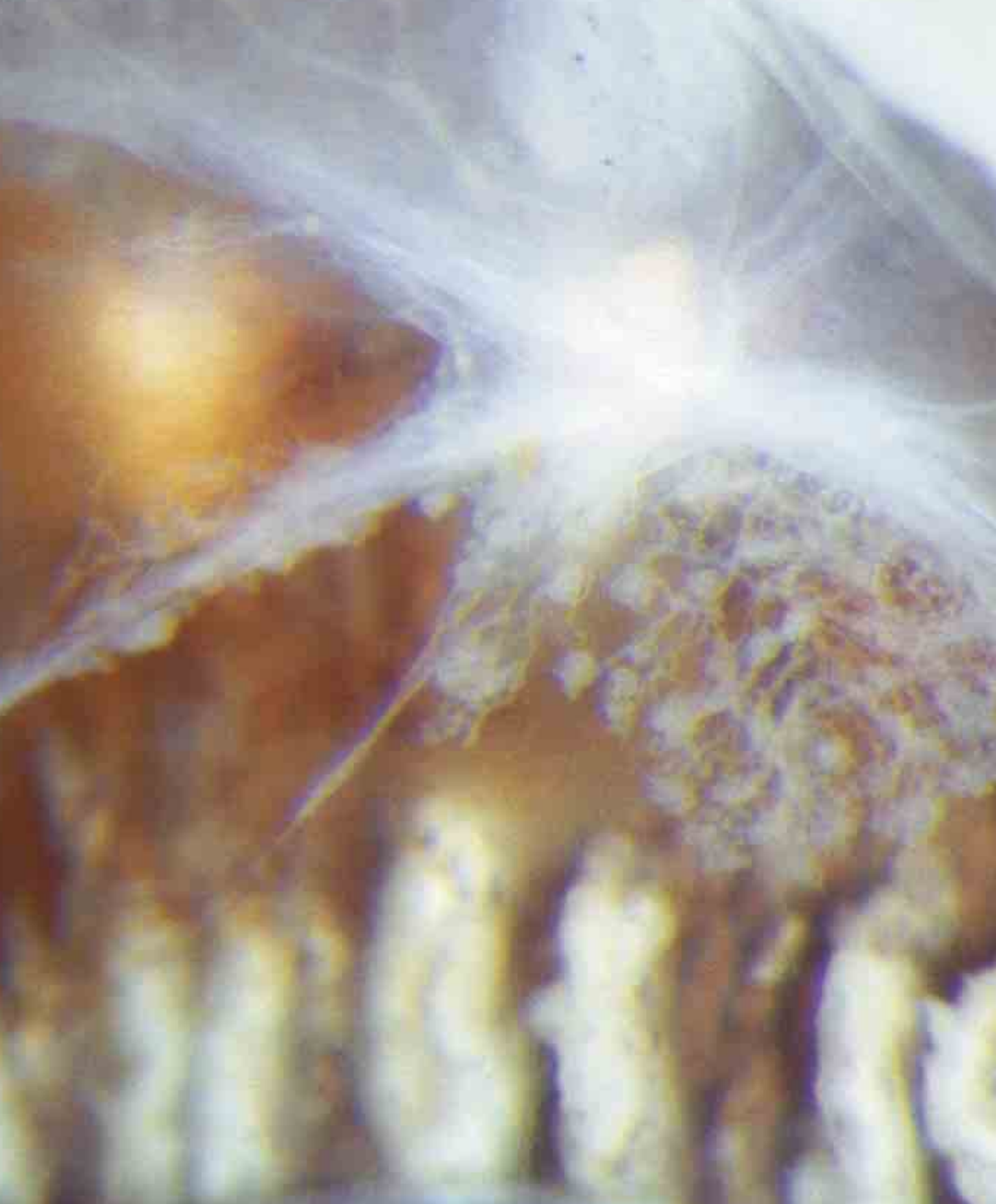
30 **Status after perforating injury (autopsy case):**
In the lower temporal periphery, a rusty metallic splinter 1 mm in thickness and 3 mm in length is embedded in fibrous tissue on the posterior pars plana.

31 **Total coloboma at the top after iridencleisis:**
Abundant pigment seeding into the space of Garnier.



- 32 **Myopic pigment epithelial degeneration:** The peripheral fundus between the ora and the equator exhibits a broad band of pigment irregularities. The posterior border is polycyclic.
- 33 **Pigment epithelial degeneration in ‘malignant’ myopia:** At the peripheral fundus there is a circular wreath of small nodules of pigment epithelial hyperplasia and individual cobblestone-like areas of atrophy. The retina shows advanced thinning and atrophy.
- 34 **Figure 33 in more detail:** Next to the small nodules of pigment epithelial hyperplasia there is advanced degeneratio linearis vasculosa (linear vascular degeneration). A peripheral cystoid retinal degeneration is missing.
- 35 **Progressing phthisis:** Epiciliary notch after destruction of the pars plana and plicata, ossified ‘Ringschwiele’, and funnel-shaped retinal detachment.





Surgery-Related Lesions

Introduction

Many enucleated globes reveal evidence of previous surgical procedures. They can also show sequelae in the area of the fundus extremus.

General Complications

Hemorrhage into the vitreous base, hemorrhage and pigment dispersion into the orbicular space, diffuse and cystic epithelial ingrowth, and changes of the vitreous base are considered as general complications.

Procedures causing destruction, collapse, shrinkage, and traction of the vitreous can lead to retinal tears and retinal detachment of the fundus extremus.

Specific Complications

(1) Cataract Surgery

Traction by callous secondary cataract formation or vitreous alterations after extracapsular cataract extraction can have an impact on the fundus extremus. Traction towards the shrinking lens remnants can pull the vitreous base gradually anteriorly and inward, eventually causing an ora tear, tears at vitreoretinal adhesions, serous detachment of the retina and pars plana and/or elongation with (pro)traction of the pre-retinal epithelium [1, pp. 150–156].

(2) Cyclodestruction by Cryocoagulation and Thermal Coagulation

The pars plicata is coagulated by application of heat or cooling energy through the sclera.

Frequently, the lesions are placed too far backwards over the pars plana. Histologically, one can identify a circumscribed necrosis of the sclera and the nonpigmented ciliary epithelium. As a consequence, there is scar formation and deposition of pigment clods by proliferation of the surviving pigment epithelium. The choroid and interstitial tissue become fibrotic. The vitreous is firmly attached to the scar, and the zonules are degenerated.

(3) Sclerotomy and Pars plana Vitrectomy

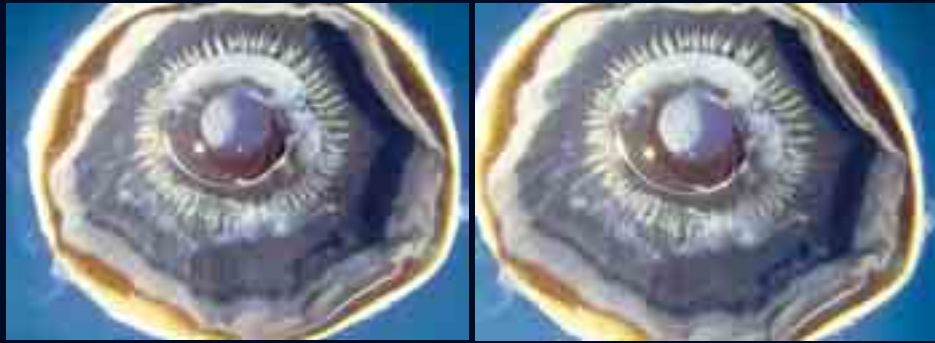
Lesions at the pars plana sclerotomy site heal with fibrovascular tissue proliferation that fills in the defect in the pars plana with little or no fibrous tissue ingrowth. After pars plana vitrectomy, there is occasionally some fibrous tissue proliferation at the inner aspect of the sclerotomy towards the lumen of the globe [11]. Together with a pars plana vitrectomy, a silicone oil tamponade can be performed. Residual intraocular silicone oil can, according to Eckhardt et al. [12], migrate into the retina where it can be found as vacuoles. It is also found within individual macrophages or as larger vacuoles surrounded by multiple macrophages in epiretinal membranes, vitrectomy scars, and within other scar tissue. Later, a cataract can develop.

- 1 **Status after extracapsular cataract extraction:** Only an upper peripheral ring-shaped bulge remains of the lens. Within the capsular bag, there is a posterior chamber lens with its lower haptic visible in the sulcus. The upper haptic cannot be seen. A fan-shaped fibrotic secondary cataract extends from the 12 o'clock position across the ring-shaped bulge towards the edge of the optic.

- 2 **Detail of the pars plana demonstrated in figure 1:** The lower lens capsule is slightly broadened and covers the apex of the ciliary processes. Disseminated onto the pars plana, clusters of cortex remnants and 'frog spawn' are visible.

- 3 **Status after dart injury, blind from secondary glaucoma:** At the site of the lens there is a dense callous scar, partially interspersed with a granular mass. Fibrous extensions of this scar extend up to the anterior border of the ciliary processes. The latter carry nodular hyperplastic excrescences of the ciliary epithelium.

- 4 **Status after extracapsular cataract extraction in a patient with Refsum's disease:** Partially calcified equatorial remnants of the lens are located inferiorly. A membrane formed by condensed vitreous and remnants of the lens capsule is traversing the center of the ring-shaped bulge; it is partially granular due to calcification and overgrown by melanocytes. The pupillary margin and iris pigment epithelium are damaged.

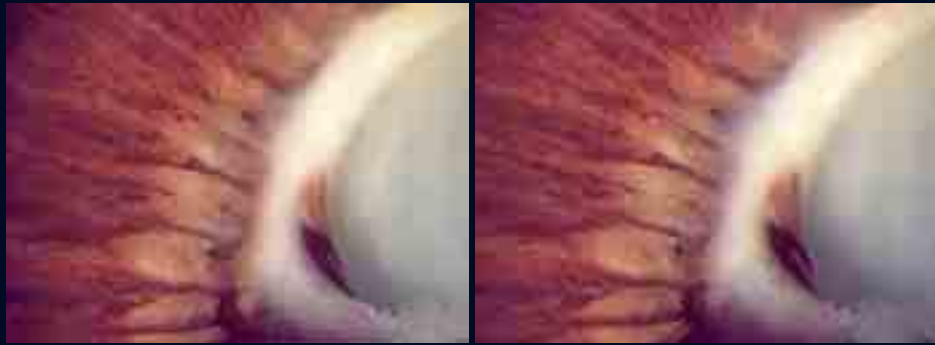


5 **Status after intracapsular cataract extraction:**
Callous fibrous scar at the upper pars plicata of the ciliary body in the region of the previous surgery site.

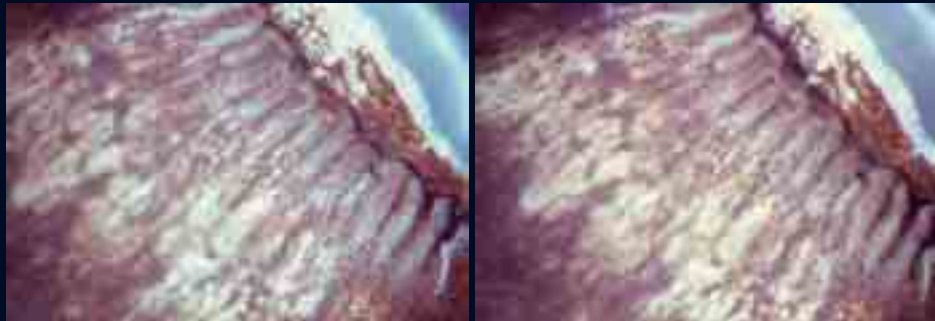
6 **Status after intracapsular cataract extraction:**
View from above into the area of the sulcus.

7 **Status after intracapsular cataract extraction:**
Ciliary processes of the third order. Adjacent, there is a nodular hyperplasia of the ciliary epithelium on the anterior pars plana. Pigment seeding onto the posterior pars plana.

8 **Status after extracapsular cataract extraction:** There is a crescent-shaped dissemination of melanin debris across the posterior pars plana. Within this material, there are a few Wedl cells.



- 9 **Status after recent cataract surgery:** There is a thin crescent of blood in the lower recess of the space of Garnier.
- 10 **Status after intracapsular cataract extraction 10 days earlier:** On the lower pars plana there is blood within the recess of the space of Garnier, partially accumulating anterior to the peaks of pars plana cysts. These are present in fairly large dimensions up to the pars plicata.
- 11 **Epithelial ingrowth after cataract surgery 8 weeks earlier:** A thin grey-whitish membrane, arising from the chamber angle, covers the pars plicata of the ciliary body.
- 12 **Status after cyclocryocoagulation for secondary glaucoma in chronic uveitis:** Temporally, there are mottled scars (coagulation scars) astride the ora and over the pars plana.



13 **Status after cyclocryocoagulation for ischemic central retinal vein occlusion:** On the upper pars plana, a polycyclic atrophic scar extends over 3 hours.

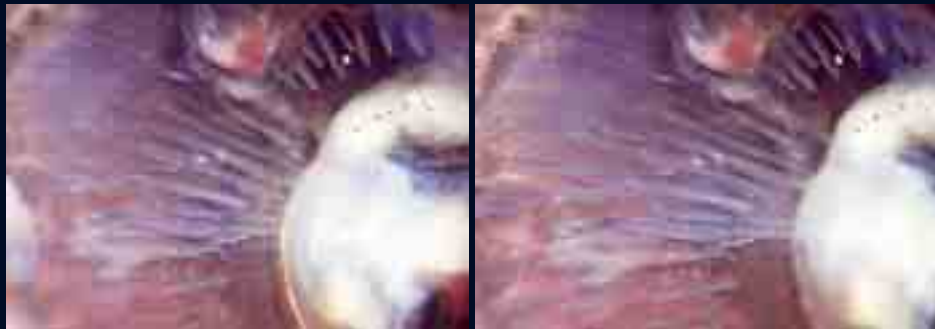
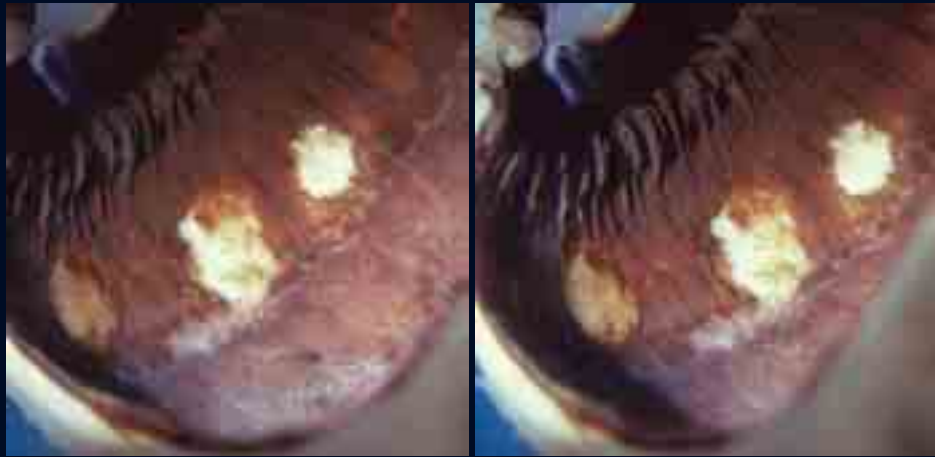
14 **Status after cyclocryocoagulation for secondary glaucoma after central retinal vein occlusion:** At the nasal-horizontal side, the ciliary processes appear atrophic. In the same sector there is a fibrous membrane covering the posterior iris surface. The lens is artificially subluxated temporal and posterior.

15 **Status after cyclodiathermy in Cogan-Reese syndrome:** There is an extensive atrophic scar on the peripheral pars plana and peripheral retina in the lower half of the globe.

16 **Status after cyclophoto- and cyclocryocoagulation for Fuchs heterochromic cyclitis:** Below between the 5 and 7 o'clock position at the posterior border of the ciliary processes there are several small atrophic coagulation scars. The ciliary processes appear undamaged. Adjacent, on the pars plana, there is a large roundish atrophic coagulation scar.



- 17 **Status after cyclodiathermy coagulation and intracapsular cataract extraction:** On the pars plana several atrophic scars which are round to polycyclic can be seen. Aphakia.
- 18 **Status after cyclodiathermy coagulation:** On the pars plana there are widespread areas of atrophic and hyperpigmented scars. Cataracta corticalis et nuclearis brunescens.
- 19 **Status after cyclodiathermy coagulation for absolute secondary glaucoma after central retinal vein occlusion:** At the level of the ora and pars plana, corresponding to the cyclodiathermy coagulation scars, one can see strand-like fibrous scarring of the inner ocular sheaths. On the temporal side, fan-like tufts of vascularized fibrous tissue emanate from these scars into the vitreous and extend across the pars plicata towards the lens, the latter being brown-yellow, well demarcated and cataractous.
- 20 On the temporal pars plana there is the scar of a subtle perforation (puncture?). From here a fan-like epithelial proliferation has grown into the space of Garnier.



- 21 **Status after vitrectomy and photocoagulation for diabetic retinopathy:** On the pars plana there is a vitrectomy scar. From here, a fan-like filamentous bundle extends across the pars plana towards a fairly large polycyclic coagulation scar at the ora.
- 22 On the mid pars plana in the lower nasal quadrant a vitrectomy scar spreads out over the pars plana, causing a retinal traction.
- 23 **Puncture scar of the pars plana:** A small feathery scar on the lower temporal pars plana.
- 24 **Status after two vitrectomies/biopsies for astrocytoma:** On the upper temporal pars plana there is a polycyclic vitrectomy scar with vitreous strands extending towards a duplicate vitrectomy scar temporally below.



- 25 **Status after two vitrectomies/biopsies for astrocytoma with dissemination of tumor onto a vitrectomy scar (same case as in fig. 24):** On the lower temporal pars plana there is the second smaller vitrectomy scar. A vitreous strand comprising whitish tumor nodules of variable size is drawn into this scar.
- 26 **Status after pars plana vitrectomy and extracapsular cataract extraction for proliferative diabetic retinopathy:** On the lower pars plicata and the lower pars plana there is extensive old hemorrhage. In the lower temporal quadrant over the posterior pars plana a vitrectomy scar can be seen. From here, a fine fibrous tuft extends into the space of Garnier. Lens remnants are present as a ring-shaped bulge of residual cortex. The haptic of the intraocular lens can be identified within the capsular bag.
- 27 **Same case as in figure 26:** There is an additional vitrectomy scar in the temporal upper quadrant. The retina is pulled towards the scar, and a tuft of newly formed vessels, some looking like hairpins, is growing out of the corresponding uveal scar.
- 28 **Same case as in figure 27 (detail):** The newly formed blood vessels terminate in small cellular clusters which look like precipitates. The vessels use a firm vitreous veil as a scaffold (probably anterior vitreous border).

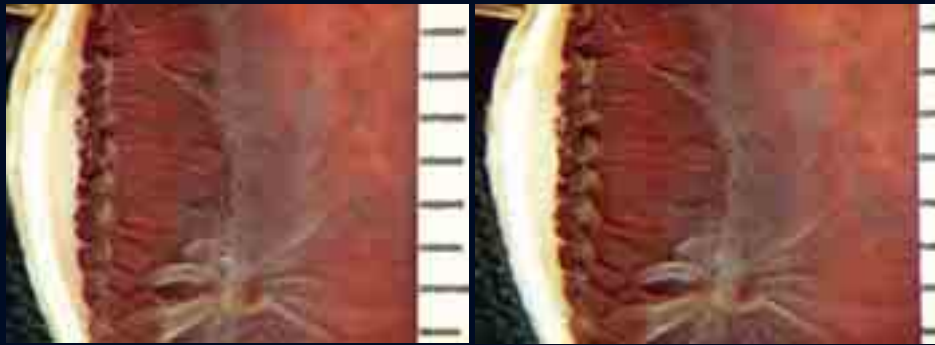


29 **Status after pars plana vitrectomy, encircling band, and silicone oil tamponade for proliferative diabetic retinopathy:** On the temporal side there is a highly elevated total retinal detachment with folds. On the temporal horizontal pars plana the feathery vitrectomy scar which caused the lappet-shaped retinal traction can be recognized.

30 **Status after vitrectomy for fungal endophthalmitis:** On the temporal pars plana there is a fine feathery vitrectomy scar with a small cone-like top and radiating vitreous adhesions.

31 **Early phthisis bulbi:** A stellate scar at the ora with meridional folds of the pars plana ciliary epithelium.

32 **Status after repeated surgery for retinal detachment and pars plana vitrectomy:** There is a prominent epiciliary scar in the nasal pars plana from which epiciliary stellate fibrous strands emanate into the vitreous.

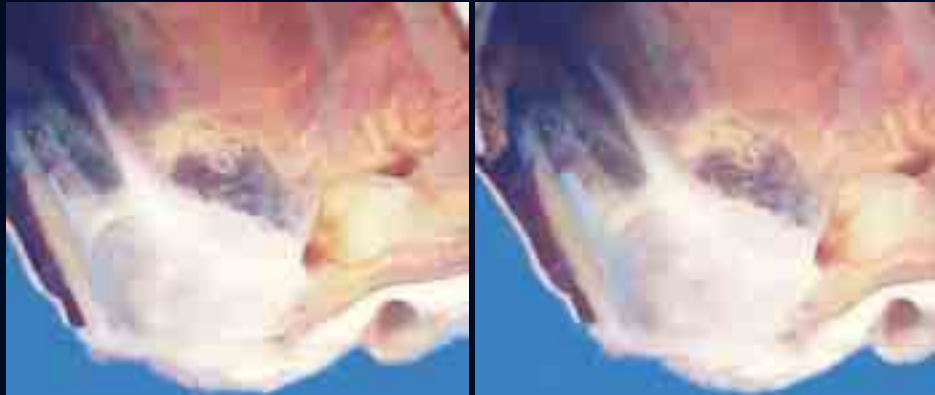
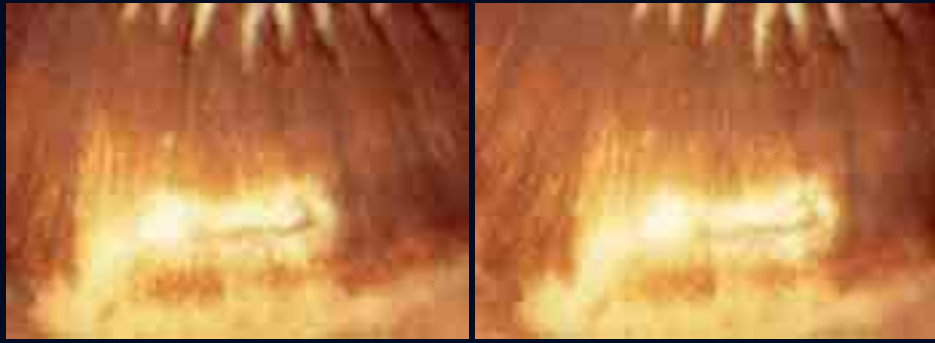


- 33 **Same case as in figure 32:** On the temporal pars plana a feathery vitrectomy scar can be seen with some residual silicone oil droplets.
- 34 **Status after corneal perforation with tractional retinal detachment, encircling band, pars plana vitrectomy and silicone oil tamponade:** The ciliary body is covered with fibrous scar tissue enclosing small oil droplets. Only stub-like remnants of the vitreous base are left behind.
- 35 **Status after pars plana vitrectomy and silicone oil tamponade:** Small emulsified silicone oil droplets are entrapped over the pars plicata.
- 36 **Status after vitrectomy for synchysis nivea:** Over the pars plana, there is a fine feathery scar with some residual white globules characteristic for this type of vitreous degeneration.



- 37 In the area of the lower pars plana there is a white scar behind a Lindner trepanation.

- 38 **Status after cyclectomy for iris melanoma:**
11–13 mm behind the limbus, the sclera is indented by an encircling band. The lower peripheral fundus reveals a snow-white surgical coloboma of the ciliary body, peripheral choroid and retina extending up to the equator. Its borders are lined by pigmentary irregularities and lipoidal changes. Fibrous strands extend from the edge of the ciliary coloboma to the corresponding sides of the iris. At the central margin of the surgical coloboma, a nodule of 4.5 mm in diameter and 2.5 mm in height can be seen; it is yellow-white and partially covered by hemorrhage.



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