Fig. 3. Immunodiffusion studies on primate β-crystallins. Gel A: 1, lemur mixed β-crystallin; 2, chimpanzee β-L; 3, calf β-L; 4, human β-L; 5, rhesus monkey β-L; 6, calf β-L. Center well contains antiserum to calf β-L.

Gel B: 1, calf β-L; 2, blank; 3, Galago senegalensis lens homogenate; 4, calf β-L; 5, Miopithecus talapoin lens homogenate; 6, Saimiri sciureus lens homogenate. Center well contains antiserum to calf β-L.

which are unique to βu is highly variable among the primates, as it is among other species.

A further point which may be significant to the area of human cataractogenesis is the presence in human βu of a higher-molecular weight SDS-polypeptide, possibly a covalently cross-linked dimer of β-crystallin subunits. It is uncertain whether this component is present in human lens in general or rather just in the ageing lens. If it is a product of ageing, it cannot be determined with certainty to be peculiar to the human lens, since the human lenses used in this study are of considerably greater relative age than those of the other species.

Further studies on primate lens proteins requiring the isolation of individual components are needed to determine the functional significance of the observations outlined above, but the consistent differences found between primate β-crystallin and that of other species may have implications for the common practice of using nonprimate models in studies of cataractogenesis.

From the Department of Pediatrics, Duke University Medical Center, Durham, N. C. This work was supported in part by NIH Grants AM 06815 and EY 01607. Submitted for publication March 3, 1976. Reprint requests: Dr. J. Samuel Zigler, Jr., Box 3854, Duke University Medical Center, Durham, North Carolina 27710. Dr. Sidbury's current address is Research Director, National Institute of Child Health and Human Development, NIH, Bethesda, Md.

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Origin of ghost cell in Coats' disease.

YOICHI TAKEI.

A morphological study of Coats' disease was performed with light and electron microscopy. The choroid was intact throughout. The pigment
epithelial cells showed proliferative changes, being thickened in two or three rows. The transitional form of ghost cell was observed in contact with the pigment epithelial layer, then migrated toward the subretinal space, and finally invaded the detached retina. It was concluded that the origin of ghost cell in Coats' disease was the pigment epithelium.

The accumulations of ghost cells in and beneath the retina were assumed to be important histological characteristics in Coats' disease.

Recently a few electron microscopic studies of Coats' disease were reported. However, the origin and role of ghost cell have been a subject of controversy: pigment epithelium, retinal histiocyte, or monocyte has been thought as its origin.

The purpose of this communication is to demonstrate the transition of pigment epithelium into ghost cell in Coats' disease.

Case history. A 4-year-old female child was referred to the hospital with a white pupil in the left eye. Fundus examination revealed an extensive retinal detachment, with a yellowish mass pushing the retina forward. There were small hemorrhages in the detached retina. The right eye was mostly normal. Because the diagnosis of retinoblastoma could not be excluded, the left eye was enucleated.

Methods. Small blocks of tissue from the retina and choroid were immersed in 2.5 per cent glutaraldehyde solution buffered with phosphate (pH 7.2) at 4°C for 1 hour, and post-fixed in 1 per cent osmium tetroxide for electron microscopy. After dehydration in graded alcohol and clearing in propylene oxide, the blocks were embedded in Epon. Thick sections were stained with toluidine blue for light microscopy. Thin sections for electron microscopy were stained with uranyl acetate and lead citrate, and were observed with JEM 100-C electron microscope.

Results. Pathologically, the specimen was diagnosed as Coats' disease because of massive eosinophilic exudate in and beneath the detached retina, dense infiltration of ghost cells in the subretinal fluid, and telangiectasis. A detailed description of the pathological findings is omitted to focus the problem to ghost cells.

Choroid. The choroid was normal throughout.

No histiocyte or monocyte was found to migrate into the pigment epithelial layer or into the subretinal space through Bruch's membrane.

Pigment epithelium. The pigment epithelium showed proliferative changes, forming a convexly protuberant apical surface (Fig. 1). In many areas the pigment epithelial cells migrated toward the subretinal space. Mushroomlike cells protruded beyond the normal height of the pigment epithelial layer. The transitional forms between the pigment epithelium and ghost cell were also found (Fig. 3). In places, the pigment epithelial cells proliferated were arranged in two to three rows (Fig. 2). Desmosomes and terminal bars were identified between adjoining epithelial cells. The cytoplasm was full of lipid droplets. Phagosomes contained lamellar inclusion bodies which were probably outer segment material. Rough-surfaced endoplasmic reticulum was well developed, whereas smooth-surfaced endoplasmic reticulum was scanty in number.

Subretinal space. Subretinal space was filled with eosinophilic homogeneous material containing cholesterin clefts. Generally the ghost cells accumulated in the vicinity of the pigment epithelial layer and detached retina, and they appeared to be floating in the subretinal space in groups. However, no types of junctional apparatus were found between the cells. The cells were large but of varying size, usually round or oval. They contained many vacuoles, less pigments, and cholesterin crystals in the cytoplasm (Fig. 4, A).

In some of the cells the cytoplasm was filled with numerous vacuoles and engulfed materials, and there were observed no normal organelles except the nucleus (Fig. 4, B).

Retina. The photoreceptor cells had been highly degraded and disappeared. Numerous ghost cells were observed in groups near the external limiting membrane; some of them migrated into the detached retina where retinal architecture was almost normal except for the slight deposition of eosinophilic exudate. However, there was no evidence of the presence of any transitional cells between the retinal cells and ghost cells.

Comments. It is generally accepted that ghost cells appear not only in Coats' disease but also in the subretinal space of Junius-Kuhnt disciform degeneration of the macula, long-standing detachment, xanthelasma, and rhegmatogenous retinal detachment. Among these, ghost cells are mostly observed beneath the detached retina in Coats' disease, and their accumulation is one of its characteristic pathological features.

Because of the morphological specificity of the cell, it is termed ghost cell, foam cell, or macrophage, or bladder cell.

As regards such ghost cells, there are many controversies about their origin even in the electron microscopic studies.

Recently Machemer and Laqua reported the phagocytic activity of the pigment epithelium, and showed the proliferation and migration of pigment epithelium transforming to macrophage. The following data support the pigment epithelial origin of ghost cell. (1) Cells separate from the proliferating pigment epithelium, showing clear transitional form between the pigment epithelium and ghost cell, and migrate toward...
Fig. 1. Pigment epithelial cells (Pig) are proliferated. Ghost cells (arrows) are floating in groups in the subretinal space (SRS). No cellular infiltration is seen in the choroid (Ch). (×200.)

Fig. 2. Proliferated pigment epithelial cells contain many vacuoles, native pigment granules, and phagosomes. (×3,500.) Inset: Light micrograph of the same region. Pig, Pigment epithelial cells; SRS, subretinal space.
Fig. 3. Pigment epithelium protrudes toward the subretinal space. (x11,000.) *Inset:* Light micrograph of transition of pigment epithelial cells (Pig) into ghost cells. SRS, subretinal space.

Fig. 4. A, Ghost cells in the subretinal fluid. Arrows indicate engulfed cholesterol crystals. (x4,000.) B, Ghost cell in its later stage is filled with a large number of vacuoles and engulfed materials. (x3,000.)
the subretinal space. (2) No cell infiltration is seen indicating invasion from the choroid into the subretinal space through the pigment epithelial layer. (3) Although a few ghost cells lie in the detached retina, their transitional form could not be found in the retina, whereas the transition of proliferating pigment epithelium into the retina was confirmed at the slightly damaged area in the peripheral retina.

From the result obtained in the present study, it can be concluded that ghost cells in Coats' disease are originated from the retinal pigment epithelial cells.

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From the Department of Ophthalmology, Tohoku University School of Medicine, Sendai-shi, Japan. Submitted for publication March 12, 1976. Reprint requests: Yoichi Takei, M.D., Department of Ophthalmology, Tohoku University School of Medicine, 1-1 Seiryu-cho, Sendai-shi, 980, Japan.

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