broken down with difficulty. It is therefore generally thought that melanin synthesis is low or absent in the adult eye, since pigmentation does not increase with progressive age.

By far the greatest amount of catechols was present in the cytoplasm, and there was very little in the large granule fraction. Since this fraction contains most melanin organelles it would hardly seem likely that the presence of dopa and 5-S-cysteinyldopa in the choroid or retina of the adult eye could reflect melanin formation. Our finding that all or almost all dopa and 5-S-cysteinyldopa is present in the cytoplasm favours the hypothesis that these pigment precursors, when located outside melanosomes, are excreted from the cell instead of forming pigment.

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Periodic Structures in the Langerhans' Cell: An Optical Diffraction Study

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Abstract. The crystalline-like structure of horizontally sectioned Langerhans cell granules has been studied by optical diffraction. Two different fixation techniques resulted in two different sets of spacings.

Key words: Langerhans cell granules; Optical diffraction

During recent years there has been an increasing interest in the Langerhans cell and its possible functions. This cell is identified in the electron microscope by a specific intracellular organelle, the so-called Birbeck granule or the Langerhans cell granule, first described by Birbeck et al. in 1961 (1). Since then several authors have studied this granule in detail (2, 8). It is described as a disc-shaped structure generally sectioned more or less perpendicularly and therefore appearing like a rod with a central linear periodicity. Occasionally the granule is sectioned horizontally and then gives the impression of a crystalline-like lattice. By using two different preparation methods we have studied the periodicities of such lattice structures by means of optical diffraction.

METHODS

Biopsies were obtained from each of two healthy, adult volunteers. One biopsy was fixed in osmium–zinc-iodide (5) and the other was fixed in glutaraldehyde and postfixed

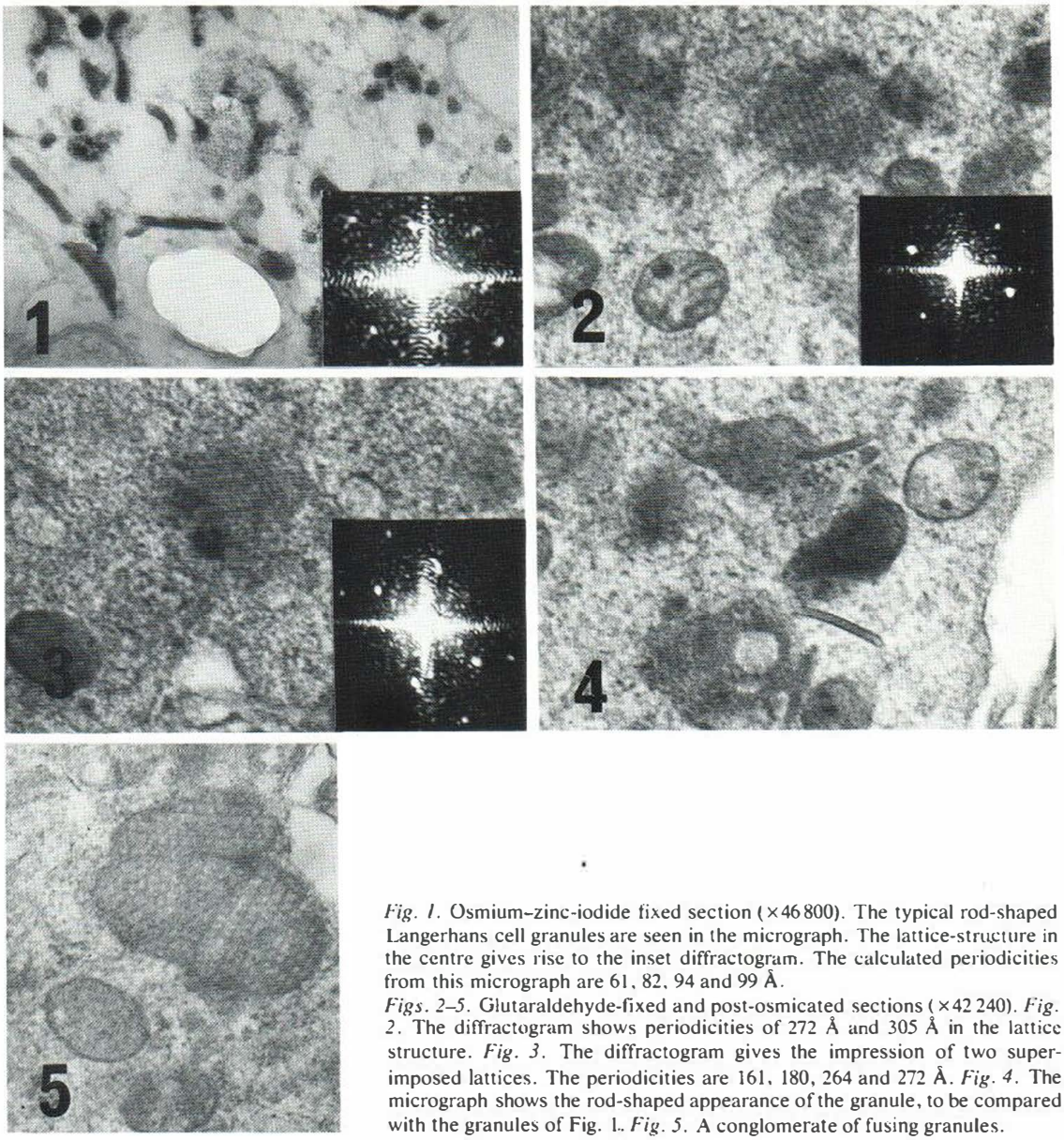

Fig. 1. Osmium–zinc-iodide fixed section (×46 800). The typical rod-shaped Langerhans cell granules are seen in the micrograph. The lattice-structure in the centre gives rise to the inset diffractogram. The calculated periodicities from this micrograph are 61, 82, 94 and 99 Å.
Figs. 2–5. Glutaraldehyde-fixed and post-osmicated sections (×42 240). *Fig. 2.* The diffractogram shows periodicities of 272 Å and 305 Å in the lattice structure. *Fig. 3.* The diffractogram gives the impression of two superimposed lattices. The periodicities are 161, 180, 264 and 272 Å. *Fig. 4.* The micrograph shows the rod-shaped appearance of the granule, to be compared with the granules of Fig. 1. *Fig. 5.* A conglomerate of fusing granules.

in osmium tetroxide. The vehicle for the fixative was distilled water in the first case and a phosphate buffer (pH 7.4) in the latter case. The specimens were prepared for electron microscopy by standard procedures. In the electron microscope goniometer the sections were rotated and tilted to compensate for oblique sectioning of the lattice structures. Recordings were made on photographic plates which then were analysed in an optical diffractometer (4).

The structure in the electron micrograph to be investigated by optical diffraction is generally positioned in a rectangular mask. The central cross of the optical diffraction patterns is thus due to the masking. Spacings in the crystalline-like lattices recorded on the electron micrographs are represented by light dots symmetrically placed in relation to the centre of the diffractogram.

RESULTS

Langerhans cells with their typical rod-shaped intracellular granules were easily identified in sections from both preparations. Structures with a lattice appearance were also seen in our sections.

Table I. *Range of periodicities in lattice-like Langerhans cell granules obtained by optical diffraction at various tilt angles*

Fixation	Range of periodicities
Glutaraldehyde and osmium tetroxide	160–451 Å
Osmium–zinc-iodide	61–107 Å

The internal structures of such horizontally sectioned granules appeared as relatively well defined points of a lattice in sections fixed by osmium–zinc-iodide (Fig. 1). The glutaraldehyde fixation with postosmication did not give rise to well defined points and the internal structure of the granule appeared as lines or as a lattice of crossing lines (Figs. 2–5). Some of the optical diffractograms from micrographs of glutaraldehyde-fixed sections also gave the impression of two superimposed lattices (Fig. 3). In some electron micrographs, granules composed of two or more fusing but separate lattices were observed (Fig. 5).

The optical diffraction analyses were performed on the different lattices observed with the two preparation methods. These lattices produced two different sets of periodicities in the optical diffractograms.

Table I summarizes our findings on the periodicity of the Langerhans cell granule. The measurements were made with the sections tilted at seven different angles. The periodicity varied according to the different tilting positions. Examples of obtained periodicity data at different tilt angle are seen in Figs. 1–3.

DISCUSSION

Tusque and Pradal (7) as well as Wolff (5) have summarized earlier published data on the periodicity of the rod-shaped Langerhans cell granule. These data were, however, all obtained with osmium tetroxide fixation. Our findings with the osmium–zinc-iodide fixation correspond closely to these data and to the results obtained with the osmium–zinc-iodide method published by Niebauer et al. (5). However, concerning normal human skin we have not found any published data on periodicities obtained with the combined glutaraldehyde and osmium tetroxide fixation.

Langerhans cell granules from Histiocytosis X lesions have also been studied with respect to the periodicities. Caputo et al. (3) have used glycerol-treated, glutaraldehyde-fixed specimens in combination with the freeze-fracture technique. They applied optical diffraction to micrographs of freeze-fracture replicas in order to calculate the periodicity. Their results are in overall agreement with these obtained from both our preparation techniques (Table II). These data were also obtained from lattice structures. Tarnowski and Hashimoto (6) have given data on the periodicity in rod-shaped granules from a case of the Letterer-Siwe disease obtained with glutaraldehyde fixation and post-osmication (Table II). To our knowledge, no reports have been published on periodic structures within the Langerhans cell, other than those on the Langerhans cell granule. At present it appears most probable that the different periodicities we have recorded result from the two different preparation methods rather than emanating from two different structures. We have however not studied any case of Histiocytosis X. If the same lattice structure has two (or more) different staining patterns we may assume that different functional groups are involved. The importance of controlled fixation parameters in studies of crystalline paracrystalline intracellular structures is stressed by our findings.

Work is now in progress in our laboratory to collect more information on the ultrastructure of the Langerhans cell granule, with the ultimate aim of a three-dimensional reconstruction of the structure.

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Table II. *Previously published data on the Langerhans cell granule periodicities*

Author	Fixation	Periodicity (Å)
Breathnach (3) 1965	Osmium tetroxide	90
Tarnowski and Hashimoto (6) 1967	Glutaraldehyde and osmium tetroxide	90
Wolff (9) 1967	Osmium tetroxide	50–60
Niebauer et al. (6) 1969	Osmium–zinc-iodide	90
Caputo et al. (4) 1976	Freeze-fracture	67; 156
	Osmium tetroxide	67; 82

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Dermatological Study of 47,XYY Males

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Abstract. Dermatological features of five 47,XYY males are presented. Port-wine stains were observed in 2 cases. As the incidence of port-wine stains among the general population is believed to be 0.5%, this result would seem to indicate that the association of 47,XYY males with port-wine stains is more than a coincidence, though the survey of previous studies failed to reveal any 47,XYY cases with port-wine stains.

A male with 47,XYY karyotype was first described by Sandberg et al. (2). Though more than 370 cases have been reported since then, no characteristic features of those males have been detected, except for high stature. Little is known about the dermatological features in 47,XYY males, the available data being limited to isolated case reports or reports involving only a few subjects—apart from the study of Voorhees et al. (3), who found an unusually early onset of acne (at 3 years of age) in 47,XYY males. They later (4) suggested that 47,XYY males often have nodulocystic acne, an observation based on a survey of patients with nodulocystic acne. They found 47,XYY males eleven times more frequently than expected.

In this study, the dermatological features of 5 47,XYY cases are described and those in a previous study are reviewed.

Table I. *Dermatological features of 47,XYY males in the literature*

Numbers indicate cases with positive findings

Acne	26
Varicose vein/ulceration	14
Dry skin	2
Hyperhidrosis	2
Wrinkly face	2
Baldness	2

Other (pigmented skin, neurodermatitis, telangiectasia, cafe-au-lait patch, tinea pedis, sebaceous adenomata, white macula, psoriasis, nevus)

MATERIALS AND METHODS

The material consisted of 5 47,XYY males found among juvenile delinquents (1). The age of the subjects ranged from 16 to 19 years. The parents of Case 1 were first cousins once removed. Those of Case 3 were first cousins. The others were unrelated. All cases were examined at dermatological interview and by inspection by one of the authors (H. N.).

In addition, dermatological features of 47,XYY subjects in previous reports were surveyed based on the compilation data in our laboratory (data available on request).

RESULTS

Three patients (Cases 3, 4, 5) displayed a mild form of acne vulgaris which emerged in adolescence. No patient showed nodulocystic acne. Port-wine stains were observed in 2 cases (Cases 3, 4). The port-wine stain of Case 3 was on the upper abdomen, 4.0×7.0 cm in size (Fig. 1). The port-wine stains of case 4 consisted of a relatively large patch (2.0×1.5 cm) and some small macules (0.5×0.5 cm) on the right forearm (Fig. 2). Another case (Case 5)