Congenital hypertrophy of the retinal pigment epithelium
Group pigmentation; bear tracks

DESCRIPTION
Congenital hypertrophy of the retinal pigment epithelium (CHRPE – ‘chirpy’) is a benign condition where the pigmented lesions may be unilateral or bilateral, solitary or multifocal, with the latter sometimes being in the form of smaller patches that are grouped together, giving the impression of animal paw-prints (bear tracks) in the fundus. Lesions of an atypical appearance may be a marker for familial adenomatous polyposis (FAP), most commonly in Gardner’s syndrome, an autosomal dominant condition where the colon polyps have a 100 per cent chance of becoming malignant. In CHRPE, the retinal pigment epithelial (RPE) cells are larger and contain more melanin granules than normal. Adenocarcinoma developing from CHRPE has been reported but is considered a rare occurrence.

SYMPTOMS
There are no symptoms associated with CHRPE.

SIGNS
A typical CHRPE is flat and black or grey, with a sharply defined border, contrasting with the slate-grey, ill-defined appearance of a choroidal naevus. They can range from very small to many disc diameters in size. Frequently, a depigmented halo lies just inside the edge of the lesion. Further depigmented areas or lacunae may develop within the lesion with time. This is sometimes referred to as a sunburst effect. Some CHRPEs are conspicuously lacking in pigment, just having a pigmented edge. The multifocal, so-called bear-track form of CHRPE is often confined to a specific area within the fundus. CHRPE that are associated with FAP are described as atypical, being oval or even fishtail-like in shape, bilateral, more isolated in position rather than grouped together and variable in size up to approximately a disc diameter. CHRPE block choroidal fluorescence on fluorescein angiography.

PREVALENCE
Multifocal CHRPE are fairly common and occur in about 1 per cent of the population. Solitary lesions in one eye are less common.

The full series of these articles will be available in the book Posterior Eye Disease and Glaucoma A-Z by Bruce AS, O’Day J, McKay D and Swann P. £39.99 For further information click on the Bookstore at www.opticianonline.net.

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