# Choroidal folds

# Dobras de coroide

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## **ABSTRACT**

Choroidal folds is considered the most prevalent funduscopic finding in cases of orbital tumors. They are ripples in the retinal pigment epithelium, Bruch's membrane, the inner portion of the choriocapillaris and in some cases, may affect the neurosensory retina, and then called chorioretinal folds. Several ocular and systemic conditions are associated with the finding and must be properly investigated and, if necessary, promptly treated. In this review we discuss the general aspects of choroidal folds, emphasizing their characteristic features in the following ophthalmological imaging tests: Retinography, fundus autofluorescence, fluorescein angiography and optical coherence tomography.

Keywords: Choroid; Indocyanine green; Fluorescein angiography; Tomography; Optical coherence tomography; Retina

### **R**ESUMO

Dobras de coroide é considerado o achado fundoscópico mais prevalente nos casos de tumor orbitário. São ondulações no epitélio pigmentado da retina, membrana de Bruch, porção interna da coriocapilar e que, em alguns casos, podem acometer a retina neurossensorial, sendo então chamadas de dobras coriorretinianas. Diversas condições, oculares e sistêmicas, cursam com dobras de coroide e devem ser corretamente investigadas e, caso necessário, prontamente tratadas. Nesta revisão iremos abordar os aspectos gerais das dobras de coroide, enfatizando suas características nos seguintes exames de imagem: Retinografia, autofluorescência, angiofluoresceínografia e tomografia de coerência óptica.

Descritores: Coroide; Indocianina verde; Angiofluoresceinografia; Tomografia; Tomografia de coerência óptica; Retina

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#### Introduction

horoidal folds are grooves in the retinal pigment epithelium (RPE), Bruch's membrane, the inner portion of the choriocapillaris and, in some instances, the neurosensory retina, in which case they are called chorioretinal folds<sup>(1-3)</sup>.

They were first described by Nettleship in 1884 as "peculiar choroidal lines" in a patient with papilledema secondary to an orbital mass lesion<sup>(1,4,5)</sup>.

#### **Clinical Aspects**

Clinically, the folds appear as linear images of alternating light and dark colour. The uppermost portion of a fold is called the peak and corresponds to the lighter region. The darker region corresponds to the depressed portion of the fold and is called the valley<sup>(4,5)</sup>. Initially they are thin and mild, similar to arrows or blood vessels<sup>(4)</sup>, and may be difficult to notice on ophthalmoscopy<sup>(1)</sup>. Over time they become larger and more pigmented, which

facilitates identification(1,4).

Choroidal folds are not an unusual finding on fundus examination<sup>(5)</sup> and are the most prevalent finding in patients with orbital tumour<sup>(6)</sup>. They are usually asymptomatic but may cause visual disturbances such as hyperopia, astigmatism, and metamorphopsia<sup>(1,2,5)</sup>. Such symptoms are most evident in patients with recent folds of sudden onset<sup>(1,2)</sup>.

Patients with folds caused by intraconal orbital tumours typically have hyperopia, while astigmatism is more common in those with extraconal tumours (6).

#### Histopathogy

Histopathological examination of choroidal folds shows grooves in the inner choriocapillaris, Bruch's membrane, and RPE. The RPE has normal features in the peaks and signs of thickening in the valleys. A study based on histopathology reported that vascular engorgement, haemorrhage, inflammation and compression of adjacent scleral tissue were the most common causes of thickening of the choroid in patients with choroidal folds<sup>(7)</sup>.

#### Ophthalmic examination and imaging

During ophthalmic examination, the best way to visualise choroidal folds is through biomicroscopy using a lens to examine the fundus, positioning the slit in an area adjacent to the one under study (the retroillumination technique)<sup>(1,10)</sup>. Using the slit perpendicular to the orientation of the folds and a green light filter also facilitates visualisation<sup>(4)</sup>. Retinal folds are the main differential diagnosis and are usually caused by epiretinal membrane<sup>(1,4)</sup>.

Ophthalmic imaging tests are helpful and present characteristic findings. Colour retinography, especially with a green filter, highlights the folds and is of great value in detecting and monitoring the condition (Figure 1 A-D)<sup>(4,9)</sup>.

Fluorescein angiography (FA) typically shows alternating hyper- and hypofluorescent lines, which are visible from the early stages and disappear in later stages without impregnation or

Table 1
Conditions that can cause choroidal folds.

Ocular Causes		
Papilloedema <sup>(2,4,5,7-9,17)</sup>	Choroidal detachment (4,7,8)	
Acquired hiperopya (1,3-5,7,8,15)	Uveal effusion syndrome (7)	
Congenita <sup>(5,7)</sup>	Venous occlusion (7)	
Retinal detachment <sup>(4,7,8)</sup>	Posterior pole staphyloma (1)	
Hypotonia <sup>(1-5,7-9,17)</sup>	Central serous retinopathy (4,5,8)	
Post-traumatic <sup>(1,4,7,8)</sup>	Microphthalmia (4)	
Choroidal tumour (1,4,5,7,8,17)	Optic disc drusen (4)	
Scleritis (1,4,5,7,8,17)	Angioid streaks (4,8)	
Uvetis (4,7,8,22)	Optic neuritis <sup>(1,2,4)</sup>	
Choroiditis (4,5,7,22)	Chorioretinal scar (8)	
Papillitis (4,7,8)	After surgery for retinal detachment by scleral introflexion (1,4,5,7,9)	
Choroidal neovascularisation (1,3-5,7,21)	Orbital causes	
Thyroid ophthalmopathy (4,7,8)	Orbital inflammatory pseudotumour (1,5,7-9)	
Orbital tumour (1,2,4,5,7-9,17)	Orbital cellulitis (1,4,5,7-9)	
Mucocele <sup>(8,18-20)</sup>	Postoperative orbital oedema <sup>(7)</sup>	
	Other causes	
Sinusitis(1,4,7,8)	Drug-induced <sup>(1)</sup>	
Carotid-cavernous fistula <sup>(4,7)</sup>	Parasellar tumour (7,17)	

Table 2

Correlation between the pattern of orientation of choroidal folds and possible diagnosis<sup>(1,4)</sup>

Padrão de orientação das dobras	Possível causa
Radial from the optic nerve (NO)	Intraconal tomour
Concentric with the convex side toward the optic nerve	Extraconal tumour
Linear, temporal to the optic nerve	Hypotonia
Radial, with the centre in any portion of the retina	Choroidal neovascularisation

leakage. Hyperfluorescent lines correspond to peaks and hypofluorescent lines correspond to valleys (Figure 1 E and F).  $^{(2,8,11)}$ 

Autofluorescence (AF) also shows a characteristic pattern. Alternating hyper- and hypofluorescent lines can be seen, however they are inverted in relation to FA. The hyperfluorescent lines in FA correspond to the hypofluorescent in AF and viceversa. This is because of the higher concentrations of RPE cells in valleys, resulting in larger amount of lipofuscin. AF is inferior to FA in showing choroidal folds, but it has the advantage of being a non-invasive test (Figure 1 G and H).<sup>(3)</sup>

The pattern shown in indocyanine green angiography (IGV) is not as characteristic as AGF and may display a smaller number of hyper- and hypofluorescent lines or a similar number of larger lines. The pattern may also change depending on the cause of choroidal folds. IGV is a useful method for studying the choroidal vascular structure and therefore plays an important role in the differential diagnosis and management of choroidal folds. (12)

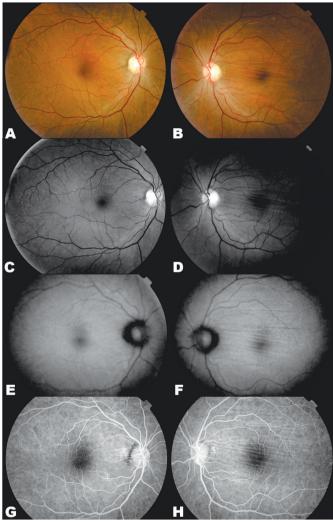
Ultrasound imaging is very helpful in determining the cause of choroidal folds, and findings can vary depending on the aetiology. The most common findings include increased subarachnoid space, flattened posterior pole, and increased thickness of the retina and choroid layer<sup>(4,5,13)</sup>.

In optical coherence tomography (OCT), folds in the hyperreflective line formed by the inner portion of the choriocapillaris, Bruch's membrane, and RPE are easily identified. In some cases the neurosensory retina remains flat over these folds, while in others it is folded (chorioretinal folds) in a pattern similar to the underlying choroid (Figure 2A and B). In a case series with 8 patients a typical pattern of vitreous attachment was noted in patients with chorioretinal folds. There was vitreous detachment in regions corresponding to valleys, while it remained positioned normally in the apical portions of the folds. (2)

Pituitary tumour associated with hydrocephalus causing increased intracranial pressure is described in the literature and is an uncommon condition. (14) Computed tomography and magnetic resonance imaging of the brain and orbits are essential to determine the presence of mass lesions and/or signs of intracranial hypertension. These tests can show signs such as a flattened posterior pole and an enlarged optic nerve sheath. (9,12,15) The enlarged optic nerve sheath at its distal portion is believed to be the cause of the flattened posterior pole. (16)

#### **Finail considerations**

Choroidal folds tend to disappear after their cause is treated successfully, but the time until total remission is quite variable and may last from a few months to a few years, and they may become permanent in some cases. In a study on choroidal folds due to orbital tumours approximately 82% of patients still had choroidal folds three months after successful treatment.



**Figure 1:** Images of a 51-year-old female with bilateral choroidal folds secondary to frontal mucocele. A and B: Simple retinography. C and D: Retinography with a green light filter. E and F: Autofluorescence. G and H: Fluorescein angiography.

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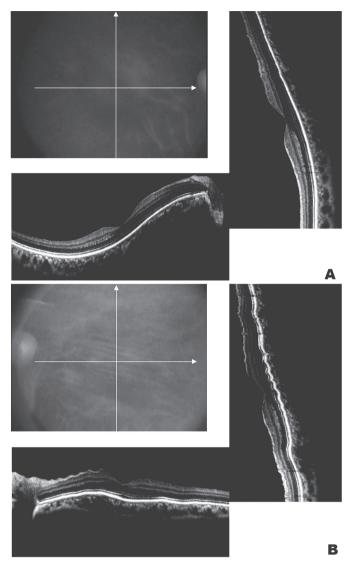


Figure 2: Optical coherence tomography of both eyes of the same patient. Perpendicular cuts through the centre of the fovea in an orientation similar to fundus examination. Figure A shows the right eve and Figure B shows the left eye

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