

# Retinopathy in patient with AC hemoglobinopathy

## *Retinopatia em pacientes com hemoglobinopatia AC*

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### **ABSTRACT**

*Hemoglobin C is the second most frequent Hb variant in Brazil and the world. Hemoglobin C trait is described as a benign and asymptomatic condition. There is little information in the literature about the association of retinal vascular disease and the presence of hemoglobin AC, being this information restricted to a few case reports. This case report describes a 26-year-old female patient with hemoglobin C trait. She presents areas of non-perfusion and arteriovenous shunts in the retinal temporal periphery of the left eye, like changes in Goldberg's stage II of proliferative sickle retinopathy. After three years of follow-up, the patient exhibits the same the alteration in right eye as well.*

**Keywords:** Hemoglobin C disease; Retinal diseases; Sickle cell disorders

### **RESUMO**

A hemoglobina C é a segunda variante de hemoglobina mais comum no Brasil e no mundo. O traço C é descrito como uma condição benigna e assintomática. Há pouca informação na literatura sobre a associação de doença vascular retiniana e a presença de hemoglobina AC, sendo esta informação restrita a alguns poucos relatos de casos. Este relato de caso descreve uma paciente do gênero feminino de 26 anos de idade com traço C. Ela apresenta áreas de não perfusão e shunts artério-venosos na periferia temporal da retina do olho esquerdo, similar ao estágio II de Goldberg de retinopatia proliferativa falciforme. Após três anos de acompanhamento, a paciente apresentou a mesma alteração também em olho direito.

**Descritores:** Doença da hemoglobina C; Doenças retinianas; Anemia falciforme

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## INTRODUCTION

The World Health Organization (WHO) estimates that 5% of the world population has some hemoglobinopathy.<sup>(1)</sup> Retinal peripheral vascular changes associated with hemoglobinopathies are well documented in the presence of hemoglobin S (Hb S), both in the homozygous form (HbSS) and in heterozygous variants (HbSC and associated with beta thalassemia).<sup>(2)</sup>

The presence of hemoglobin AC (C trait) is a condition that usually presents a benign course, with few reports of its association with retinal disease.<sup>(3-5)</sup>

This case report describes findings of alterations in the retinal peripheral vascularization of a patient with hemoglobin C trait.

## CASE REPORT

A 26-year-old female patient complained of seeing a black spot in her left peripheral vision in a routine ophthalmologic consultation in April of 2015. On the exam she presented best corrected visual acuity of 20/100 in the right eye and 20/20 in the left eye due to anisometropic amblyopia. At the indirect ophthalmoscopy examination, no retinal changes were observed in the right eye; in the left eye ghost vessels, hemorrhagic points and areas of nonperfusion were observed in the temporal periphery.

After 2 weeks of the initial consultation, the patient returned with requested exams, including hemoglobin electrophoresis, which showed a hemoglobin C pattern of 37.7%. She returned four months later with a result of angiofluoresceinography (AGF), which presented areas of non-perfusion and arteriovenous shunts in the retinal temporal periphery of the left eye, similar to changes in Goldberg's stage II of proliferative sickle retinopathy.<sup>(2)</sup> (Figure 1) After three years of follow-up, it evolved with the same alteration observed by AGF in temporal periphery of the right eye.

## DISCUSSION

Hemoglobin C is the second most frequent Hb variant in Brazil and the world.<sup>(2)</sup> In a study performed in a maternity hospital in Salvador de Bahia, a prevalence of 9.8% of HbAS and 6.5% of HbAC was observed, being the higher prevalence of hemoglobin variants described in Brazil.<sup>(6)</sup> Hemoglobin C does not present with sickling of the red blood cells, but this condition can result in an increase in blood viscosity and cellular stiffness, reducing the red blood cell life.

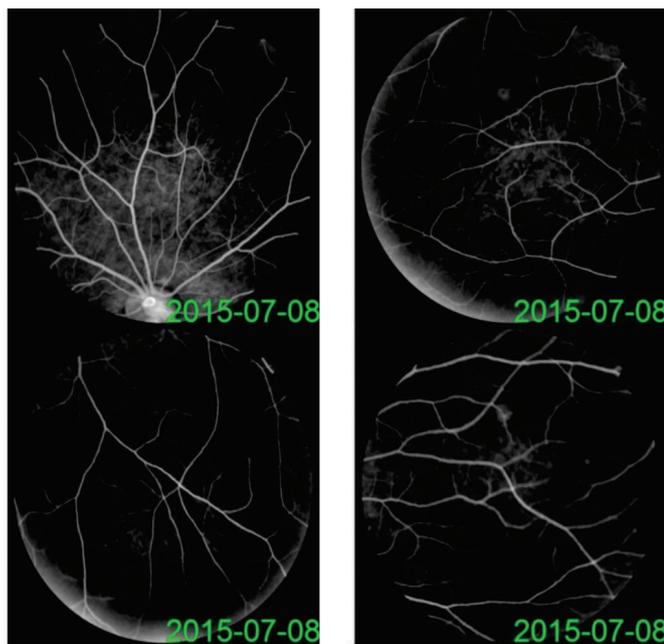
Hemoglobin C trait is described as a benign and asymptomatic condition, which presents a life expectancy compatible with normality.<sup>(7)</sup> There is little information in the literature about the association of retinal vascular disease and the presence of hemoglobin AC, being this information restricted to a few case reports.<sup>(3-5)</sup> Since it is a prevalent condition in our country, especially in the state of Bahia, further studies on this association are necessary, and it is therefore valid to consider the referral of these patients to the ophthalmologic service.

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**Figure 1:** Angiofluoresceinography of the retinal temporal periphery of the left eye.