Decreased visual acuity from optic disc drusen

P. Gili Manzanaro, J. Yangüela Rodilla, G. Rodríguez Caravaca, C. Carrasco Font, J. C. Martín Rodrigo and A. Arias Puente

Ophthalmology Unit, University Hospital Foundation Alcorcón, Madrid, Spain
Preventive Medicine Unit, University Hospital Foundation Alcorcón, Madrid, Spain

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ABSTRACT

Purpose: To evaluate the visual acuity in patients with optic disc drusen and its relationship with the existence of superficial drusen.

Methods: For a six-year period (from 1997 to 2003), fifty-five patients (100 eyes) with optic disc drusen, confirmed with B-echography, were diagnosed. According to their ophthalmoscopic appearance under 20° retinography, the drusen were classified as hidden or visible. We evaluated the best corrected visual acuity. When the visual acuity was less than 0.8, other additional causes of visual impairment were studied.

Results: The average visual acute in patients with papillary drusen was 0.82 (maximum 1.2 and minimum 0.05). Lower visual acute was found in those patients with visible drusen. Visual acute was statistically worse (p=0.016) as the number of drusen increased. The visual acute was normal in 75 cases. The decreased visual acute was exclusively secondary to drusen in 5 cases. In the other 20 patients concomitant causes of visual impairment were found.

Conclusions: A decreased visual acute secondary to drusen is unusual. When present, it is usually moderate and associated with disturbances of the peripheral visual field. There is a significant relationship between the decrease in visual acute and the number of visible drusen. When the central vision is decreased, but not the visual field, other concomitant conditions that could also affect the vision, should be ruled out.

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Introduction

The prevalence of papillary drusen has been estimated between 3.4 and 24 per 1,000 inhabitants, occur more frequently in Caucasians and exhibit a slightly higher prevalence in women.\(^1\) The age of onset is highly variable, but in most cases papillary drusen are diagnosed between the second and third decade of life in a routine ophthalmological assessment.\(^2\) Although in the initial stages said drusen are frequently asymptomatic, the visual field defects associated to drusen are common (75%) and could be significant.\(^3,4\)

The objective of this paper is to assess visual acuity (VA) alterations in patients with papillary drusen and their relationship with the presence and number of superficial drusen.

Subjects, material and methods

A retrospective transversal study was designed to assess VA in patients with papillary drusen and their relationship with the number and abundance of ophthalmoscopically visible drusen. The study was carried out in patients diagnosed with papillary drusen referred to the Ophthalmology Unit of the Alcorcón Hospital Foundation (Health Area 8 of the Community of Madrid, Spain) during the period comprised between December 1997 and December 2003. A color photo of the papilla (retinography) as well as a two-dimensional echography (eco-B) was taken of all the patients in the study to assess the optic nerve. All the tests were carried out by the same author. The echography was taken as reference criterion or gold standard for diagnosing the papillary drusen.\(^5\)

The ocular fundus images (retinographies) were taken with an eye fundus camera with telecentric optic (Zeiss FF 450 IR plus) and digital analysis archiving system Visupac 451 (version 3.2.1) (Carl Zeiss Jena GmbH, Ophthalmics Instrument Division, Jena) utilizing a 3 CCD color camera (Sony Power HAD) (resolution 786 x 576 pixels), with a 20° angle (29x) for studying the papilla. Following the Roh\(^6\) classification, we differentiated the following on the basis of the ophthalmoscopic appearance (fig. 1):

1. Hidden or deep papillary drusen: usually appearing as elevated papilla with poorly defined edges, without visible nodular images. The presence of hidden or buried papilla was confirmed with echography.

2. Visible or superficial papillary drusen: defined as yellowish nodular images having variable size, in a generally elevated papilla with poorly defined edges. The visible drusen were classified on the basis of their abundance as scarce (between 1 and 5), numerous (between 5 and 10) and very abundant (when the number exceeded 10 and occupied the entire papilla).

We recorded the Best corrected VA in the decimal scale utilizing an optotype projector Takagi CP-30 (Takagi Seiko, Co. Ltd.). Visual acuities equal to or above 0.8 were considered to be “normal”. When visual acuities were below 0.8 supplementary explorations were carried out to determine whether the papillary drusen were the only cause of reduced vision or other ocular pathologies were also present.

The study excluded patients who did not complete the two diagnostic tests (eco-B and retinography) due to opacity, poor cooperation or lack of follow-up, and those who had...
incompatible clinical records or in whom it was not possible to record the Corrected VA.

The data were registered in a data collection sheet specifically designed for the purpose. A relational database in standardized Access was designed to record all the study data.

In the study of papillary drusen characteristics, each eye was considered in isolation "case"). However, in the analysis of epidemiological factors, age, sex, history, etc., the number of patients was considered. The qualitative variables are described with their frequency distribution and compared with Pearson's $X^2$ test. The quantitative variables are described with the mean value, standard deviation (SD) and confidence intervals (CI) at 95% and compared with the $t$ for Student test when they followed normal laws. When they didn't, they were compared with the Mann-Whitney non-parametric U test. The sex qualitative variable was compared with the binomial non-parametric test. The data were analyzed with the SPSS version 12.0 statistical application.

Results

The study included 100 eyes (cases) of 55 patients with papillary drusen confirmed with echo-B. Forty-five patients (81.8%) exhibited bilateral drusen. The mean age of patients was 37.27 years (SD=20.20 years, CI at 95% of 31.81-42.74), with a minimum of 5 years and a maximum of 71 years. The distribution per sex exhibited a slight prevalence of women (58.2%, 32 women, 23 men), although without significant differences ($p=0.281$).

As regards records, 6 patients exhibited algoid striations (10.9%), 3 with elastic pseudoxantoma confirmed with pathological anatomy. A further 3 exhibited pigmentary retinosis (5.5%). At the ophthalmoscopic level, the drusen were (fig. 2):

1. Hidden or deep drusen (not ophthalmoscopically visible): 48%
2. Visible or superficial drusen: 52%.

According to abundance:

a) Visible scarce: 20%
The mean VA in eyes having papillary drusen was of 0.82 (CI 95% 0.77-0.87), with the lowest VA being of 0.05 and the highest 1.2. Acuity was normal (0.8 or better) in 75% of cases. In five cases (5%) the VA was abnormal only because of drusen and 20 cases (20%) exhibited associated visual reduction causes: 10 cases with causes unrelated to drusen, 10 with diseases or complications associated to drusen (pigmentary retinosis, angioid striations, ischemic optic neuropathy). Some cases exhibited several causes of reduced VA. Table 1 summarizes the causes of reduced VA in our series.

The five eyes (3 patients: one with unilateral drusen and two with bilateral drusen) with VA reduction exclusively due to drusen were exhaustively studied, including fluorescein angiography, computerized axial tomography, nuclear magnetic resonance and electrophysiological studies. In all the cases, they had associations with important peripheral campimetric losses (one patient with legal blindness due to concentric loss). Visual acuity values were of 0.2; 0.3; 0.5; 0.6 and 0.7.

### Table 1 – Causes of diminished visual acuity in papillary drusen

<table>
<thead>
<tr>
<th>Visual acuity reduction (number of cases)</th>
<th>Causes (number of cases)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Non-related drusen (10)*</td>
<td>Elevated refractive defects (5)</td>
</tr>
<tr>
<td>Associated diseases (7)</td>
<td></td>
</tr>
<tr>
<td>Associated complications (3)</td>
<td>Exclusively drusen (5)</td>
</tr>
<tr>
<td>Exclusively drusen (5)</td>
<td></td>
</tr>
<tr>
<td>Total number</td>
<td></td>
</tr>
</tbody>
</table>

* Some cases with several causes of visual acuity reduction. Cases=eyes; ARMD: Age Related Macular Degeneration

Comparing the VA according to the type and abundance of visible drusen, we found poorer visual acuities in patients with visible drusen, with worse results corresponding to increased amount of drusen (table 2 and fig. 3), with statistically significant differences between all the groups (p=0.016).

### Discussion

VA is usually not affected in patients with papillary drusen. VA loss exclusively due to drusen is infrequent (from 1% to 8.69%; 5% in our series). Lorentzen described a case of 91 eyes studied, Rosenberg a case of 151 eyes, and in 4 eyes of the series of 307 cases published by Mustonen. More recently, Wilkins carried out a retrospective revision of 92 eyes belonging to 56 patients with papillary drusen confirmed with echography where he described a mean VA of 1 with 8 cases (8.69%) of patients with VA under 0.8. However, Scholl described 8 eyes out of 29 with vision under 0.8. The authors

### Table 2 – Visual acuity in patients with papillary drusen

<table>
<thead>
<tr>
<th>Type of drusen</th>
<th>N%</th>
<th>Mean BCVA</th>
<th>CI 95%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hidden drusen</td>
<td>48</td>
<td>0.88</td>
<td>0.82-0.94</td>
</tr>
<tr>
<td>Visible drusen, scarce</td>
<td>20</td>
<td>0.85</td>
<td>0.74-0.96</td>
</tr>
<tr>
<td>Visible drusen, many</td>
<td>22</td>
<td>0.75</td>
<td>0.60-0.89</td>
</tr>
<tr>
<td>Visible drusen, very abundant</td>
<td>10</td>
<td>0.63</td>
<td>0.43-0.84</td>
</tr>
<tr>
<td>Total</td>
<td>100</td>
<td>0.82</td>
<td>0.77-0.87</td>
</tr>
</tbody>
</table>

Men BCVA: mean best corrected visual acuity; CI 95%: confidence interval of 95%; N%: number of cases (eyes) in percentage values.
explain these numbers due to the probable inclusion of severely affected and symptomatic patients. These studies show that the papillary drusen can cause a slight to moderate VA reduction (table 3).

However, the literature comprises publications of severe VA losses, in most cases unilateral progressive visual deterioration with peripheral campimetric losses before the central loss, associated to campimetric alterations typical of drusen in the contralateral eye. Knight13 published the case of a 40 year-old woman with bilateral drusen who exhibited a VA reduction up to amaurosis in the left eye during a three-year period. Frisen14 described a 61 year-old man with a gradual unilateral VA loss up to hand movement during a 20-year period, with bilateral campimetric alterations. In turn, Sanders15 published the case of a 63 year-old woman with unilateral progressive visual loss (VA=0.2), associated to an important concentric field loss preserving central VA, in whom other cases were excluded.

The presence of diminished VA, described between 16%9 and 28%12 of the main series on papillary drusen in 25% of our series), leads us to suspect the presence of other associated causes. The causes which could induce VA reductions in drusen patients are varied:

1. Complications associated to papillary drusen: hemorrhage, arterial and venous vascular occlusions, neovascular membranes, etc.
2. Diseases associated to drusen: pigmentary retinosis, angioid striations.
3. Other concomitant causes not related to drusen.
4. Exclusively due to drusen.

Vascular complications associated to papillary drusen can account for visual and campimetric losses. Ischemic optic neuropathies (ION) associated to papillary drusen and profusely described in the medical literature16-18 have appeared in three of our cases. Lorentzen8 stated that ION due to a vascular occlusion is the most frequent cause of VA loss in papillary drusen. The presence of small optic papilla seems particularly predisposing, supporting the theory that the drusen produce a compression of venous veins leading to an ION. Other vascular complications described as arterial retinal vessel occlusions19-21 and venous occlusions22,23 were not present in our cases. The association of subretinal neovascular membranes to papillary drusen has been described in young patients and children.24,25 Neovascularization is typically localized next to the papilla, and sometimes extends to the macula. The primary involvement of the macula is very rare.

An additional cause of vision reduction is the presence of diseases typically associated to drusen, such as pigmentary retinosis and angioid striations. In our study we have found 6 patients with angioid streaks (10.9%, 3 with elastic pseudoxantoma), 3 with pigmentary retinosis (5.5%) and none with arteriohepatic dysplasia or Alagille syndrome. These findings match those published by other authors, who report a high prevalence in papillary drusen patients of elastic pseudoxantoma between 1.4% and 3.6%1,8 (5.4% in our series), retinitis pigmentosa between 1.4%8 and 9.2%26 (5.5% in our series).

However, VA and/or campimetric loss can be due to other concomitant causes, both oculal and general, without relation to drusen. In our revision we have found five cases with severe refractive defects, four with ambylophia due to anisometropy, one corneal leukemia, one evolving cataract, two cases with Age Related Macular Degeneration (ARMD) and one macular edema. The presence of intra-cranial tumors is particularly important as its diagnostic could be delayed due to the presence of associated drusen.1,7

However, the assessment of VA in our study has shown a statistically significant relationship with the presence and abundance of ophthalmoscopically visible drusen. VA is lower in patients with visible papillary drusen and it worsens as the number thereof increases.

To conclude, diminished VA associated to papillary drusen is infrequent and when it appears it is usually moderate and nearly always associated to peripheral field alterations. VA exhibits a significant relationship with the presence and abundance of visible drusen. The loss of central VA without campimetric involvement must lead us to suspect associated causes.

### Table 3 - Visual acuity reduction in papillary drusen: bibliographic revision

<table>
<thead>
<tr>
<th>Authors</th>
<th>Number of cases</th>
<th>Diminished visual acuity</th>
<th>Only due to drusen</th>
</tr>
</thead>
<tbody>
<tr>
<td>Lorentzen8 1966</td>
<td>70 patients</td>
<td>14 (20%)</td>
<td>1 (1.42%)</td>
</tr>
<tr>
<td>Rosenberg79 1979</td>
<td>98 patients/151 eyes</td>
<td>16 (16.32%)</td>
<td>1 (1.02%)</td>
</tr>
<tr>
<td>Mustonen10 1983</td>
<td>307 eyes</td>
<td>57 (18.56%)</td>
<td>4 (1.3%)</td>
</tr>
<tr>
<td>Scholl12 1992</td>
<td>16 patients/29 eyes</td>
<td>8 (28%)</td>
<td></td>
</tr>
<tr>
<td>Wilkins11 2004</td>
<td>56 patients/92 eyes</td>
<td>8 (8.69%)</td>
<td></td>
</tr>
<tr>
<td>Gili 2007</td>
<td>55 patients/100 eyes</td>
<td>25 (25%)</td>
<td>5 (5%)</td>
</tr>
</tbody>
</table>

**References**