Case Record 6

Pigmentary Dispersion Syndrome
Pigmentary Glaucoma

January 2012

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aaron optometrists

Introduction
Referral Based on 'Intent to Treat' Criteria

The ophthalmology department and the Royal Victoria Infirmary has a document giving guidelines on when to refer suspect glaucoma; this is based on an 'intent to treat' criteria (www.newcastle-hospitals.org.uk) (Appendix 1).
Optometrists should be confident to monitor Pigmentary Dispersion Syndrome (PDS) until signs of Pigmentary Glaucoma (PG) manifest.

Defining PDS/PG for Treatment Purposes

Both PDS and PG have characteristic features; pigment deposition within the anterior and posterior segments, concave iris configuration and classic, spoke like transillumination defects of the iris (Campbell and Schertzer 1996). These authors suggest that the differential diagnosis of PDS versus PG is made purely on pressure, regardless of the status of the visual fields or optic nerves.

Migliazzo, Shaffer, Nykin and Magee (1986) propose a more functional definition; PG being diagnosed if, in conjunction with the accepted signs of PDS, glaucomatous cupping and/or visual field defects are present. This is more representative of current concepts in defining the glaucomas, which rely on disc appearances and perimetry rather than tensions (Wilson and Martone 1996) and reflects the definition of glaucoma which no longer includes the term intraocular pressure (European Glaucoma Society 2003, NICE 2009).

No latitude in either definition allows for the possibility of an alternative form of glaucoma co-existing with PDS.

February 2002

Salient information taken from electronic records

DATE: 12/2/02

Mrs Age 49
Address

Presenting Symptoms
Routine eye exam. Distance and Near vision fine – although needs more light to read. No diplopia. No HAs.

POH
Bifocal spectacles. No previous ocular surgery or treatments. Spectacles from age 15.

FOH
None.

General Health and Medications
Non-smoker. No allergies, No Hayfever
No topical, systemic Medications. General health excellent.

No previous history of general or ocular medication use or surgery.

Refraction
R -3.50/-0.50x60 (6/5) Add +2.00 N5
L -5.00/-0.75x180 (6/6) Add +2.00 N5
Phorias Dist- 3Exo Near Orthophoroc

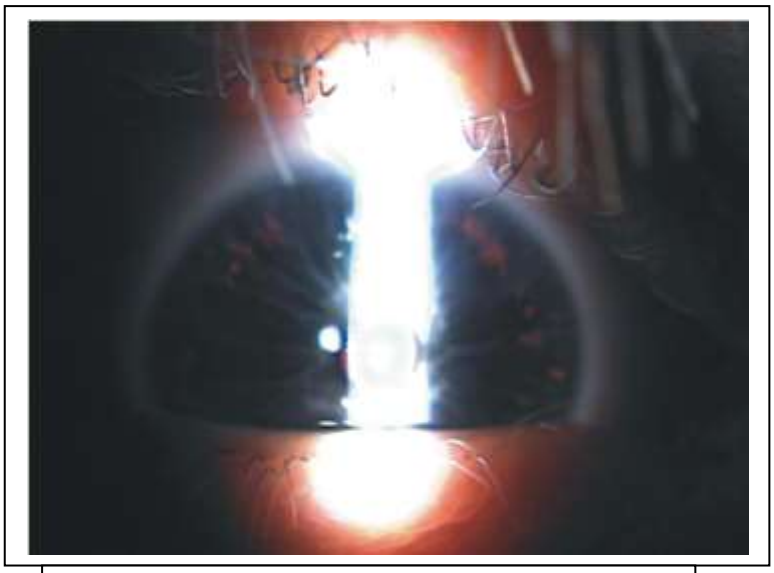
Tensions (GAT)	R 12 L 12	Glaucoma Screen
Pupils	E&A D,C& N	Full

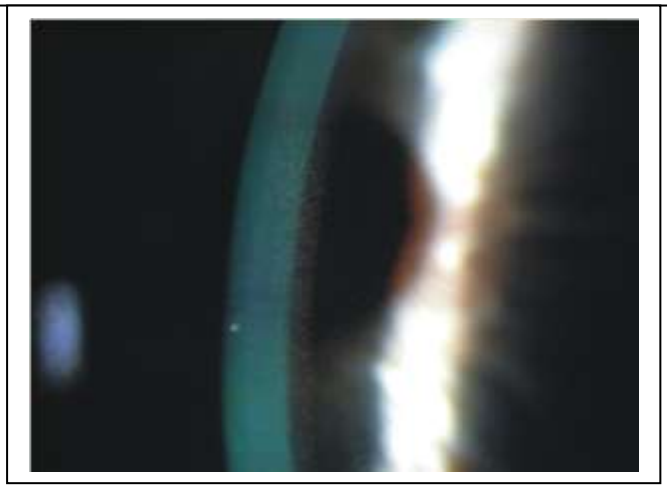
Slit Lamp
VH 4+, Concave Configuration Deep. Krukenburgs Spindle noted R&L. Spoke-like iris trans-illumination defects. Photographed

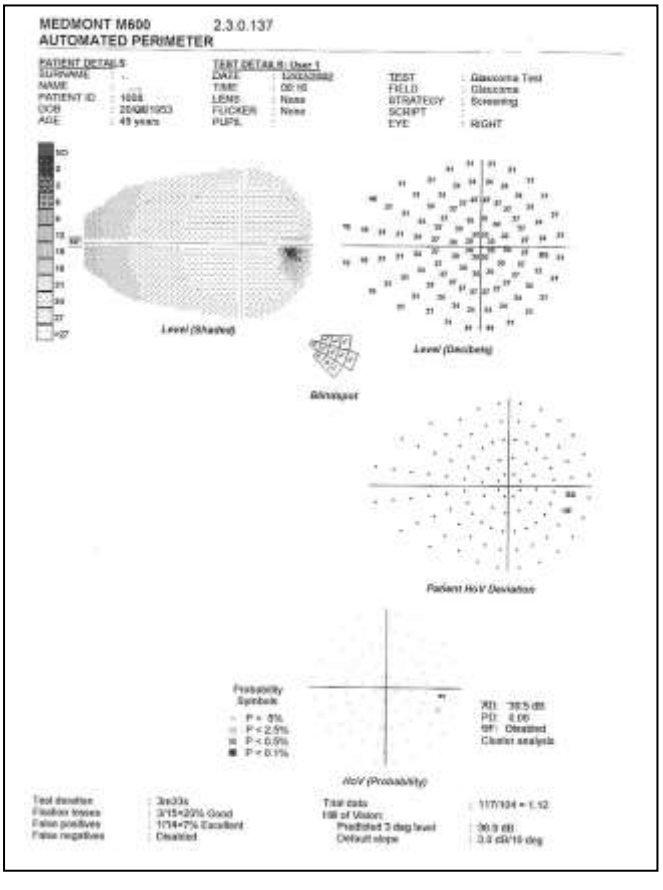
Dilated Fundsocopy (1% Tropicamide)
Nuclear Sclerosis. No Pseudo-exfoliation
Right and Left Discs VCD 0.4 Neural rims healthy and uniform (ISNT conforms) No barring, no bayoneting, No PPA

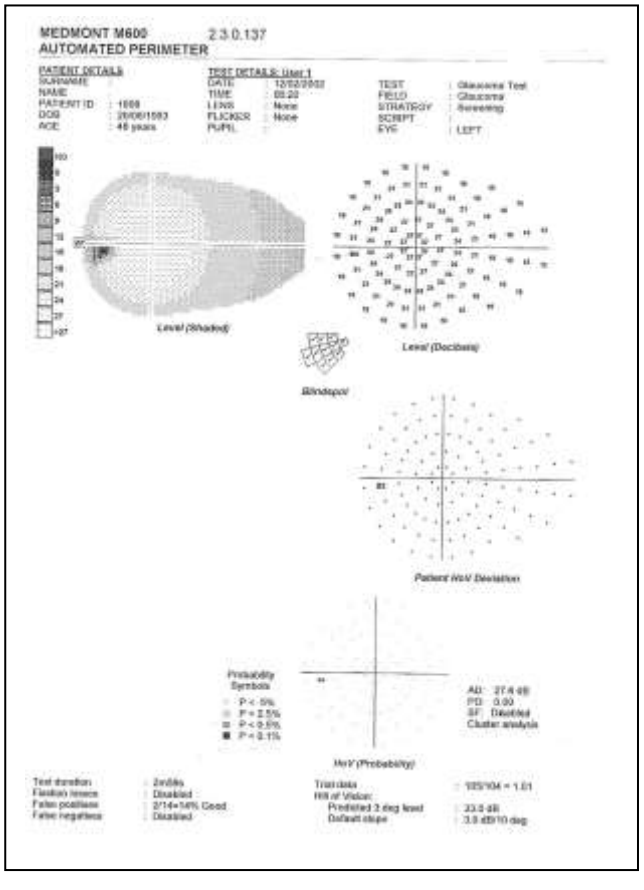
Advice and CMP
PDS diagnosed. PDS vs PG explained to Px and need to monitor stressed.

Report (information only) sent to GP and copy given to Px. Review 12/12





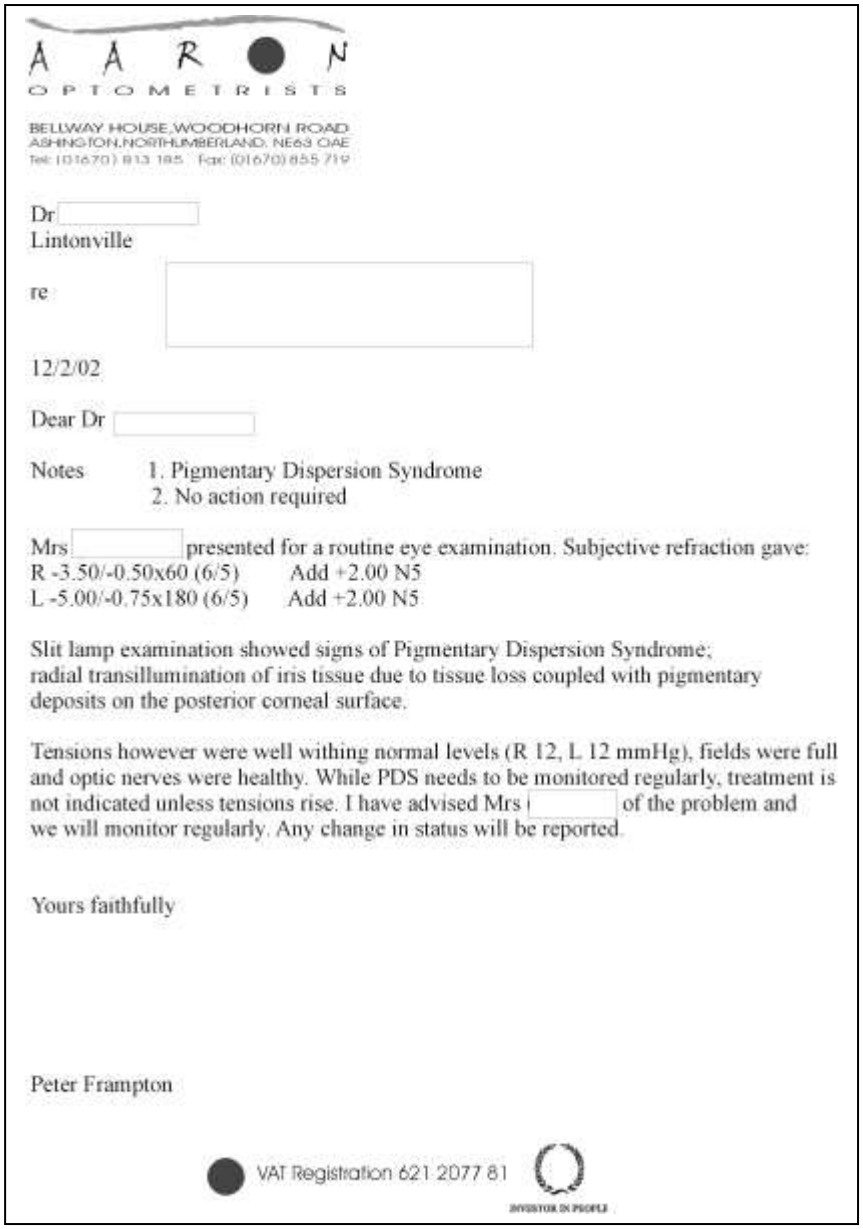




Ritch and Liebmann (1994) suggest that many cases of PDS remain undiagnosed; routine slit lamp identifying the condition. All three classic observations were identified in this blue-eyed lady, without recourse to gonioscopy. Gonioscopy allows assessment of the trabecular meshwork (TM) for depth and uniformity of pigmentation and allows observation of pigment on the posterior surface of the lens (Palmberg 1996, Campbell and Schertzer 1996).
PDS and PG are recognised to go through active (PDSa/PGa) and inactive (PDSi/PGi) stages (Campbell and Schertzer 1996, Ritch and Liebmann 1996) and the conditions have been observed to regress and ocular structures regenerate (Ritch 1982). Migliazzo et al (1986) describe the archetypal patient as a young, white, myopic male. Campbell and Schertzer (1996), remark that the disorder generally affects young adults up to 45 years of age. In the majority of cases the severity of involvement of both PDS and PG decreases in middle age when pigment liberation ceases (Ritch and Liebmann 1996), possibly due to lens growth pushing the iris forward and away from the zonules and reduced accommodation (Campbell and Schertzer 1996). These authors however, also note that women manifesting PG tend to be a decade older than their male counterparts with average age of conversion being 34 to 46 years.

At 48 years of age at presentation, this patient was an established presbyope, but Campbell and Schertzer (1996) don't exclude older patients from manifesting active pathology.
Normal discs and full fields did not suggest regressed PG and with tensions as low as 12mmHg in each eye the patient was confidently diagnosed as PDSi.
A suprathreshold strategy was conducted at this time; useful as a screening tool to improve specificity, but at the loss of sensitivity, this strategy should not have been used in this situation. Acceptable to confirm the other clinical signs of healthy disc and normal pressures, suprathreshold strategies do not give Statpac data for the monitoring of subtle progression (Anderson 1992); this set of fields could not constitute a baseline.

The decision to monitor was made, full information given to the patient who agreed with the proposed plan and a report sent to her GP.



AARON
OPTOMETRISTS

BELLWAY HOUSE, WOODHORN ROAD
ASHINGTON, NORTHUMBERLAND, NE63 0AE
Tel: (01670) 813 185 Fax: (01670) 855 719

Dr
Lintonville

re

12/2/02

Dear Dr

Notes 1. Pigmentary Dispersion Syndrome
2. No action required

Mrs presented for a routine eye examination. Subjective refraction gave:
R -3.50/-0.50x60 (6/5) Add +2.00 N5
L -5.00/-0.75x180 (6/5) Add +2.00 N5

Slit lamp examination showed signs of Pigmentary Dispersion Syndrome; radial transillumination of iris tissue due to tissue loss coupled with pigmentary deposits on the posterior corneal surface.

Tensions however were well withing normal levels (R 12, L 12 mmHg), fields were full and optic nerves were healthy. While PDS needs to be monitored regularly, treatment is not indicated unless tensions rise. I have advised Mrs of the problem and we will monitor regularly. Any change in status will be reported.

Yours faithfully

Peter Frampton

VAT Registration 621 2077 81

Annual Reviews 2003 till 2007

At the routine reviews tensions remained stable and well within normal limits (12mmHg to 14mmHg). The Royal College of Ophthalmologists (2004) advise caution when interpreting single IOP readings. Measurement error can be attributed to equipment/patient/operator variability (Chihara 2008, Royal College of Ophthalmologists 2004, Schottenstein 1996, Zeimer 1996, Whiteacre and Stein 1993, Piltz, Starita R, Miron M and Henkind 1985).

However isolated clinical findings should correlate with the overall clinical picture. Fields remained full to Fast Threshold strategies and disc appearances were normal and stable. At each annual review, findings indicated PDSi.

March 2007

Salient information taken from electronic records

DATE: 30/3/07

Mrs DOB Age 54
Address

Presenting Symptoms
Routine review for PGS. Distance and Near vision remains good. No diplopia. No HAs.

POH
Bifocal spectacles. No previous ocular surgery or treatments. Spectacles from age 15.

FOH
None.

General Health and Medications
Non-smoker. No allergies, no Hayfever
No Medications. General health excellent.

No previous history of general or ocular medication use or surgery.

Refraction

R -3.25/-0.50x65 (6/5)	Add +2.00 N5	Intermediate +1.75
L -5.00/-0.75x180 (6/6)	Add +2.00 N5	Intermediate +1.75

Phorias Dist- 3Exo Near Orthophoroc

Tensions (GAT)	R 19 L 19 @ 11.15	Glaucoma Fast Threshold
Pupils	E&A D,C& N	Full

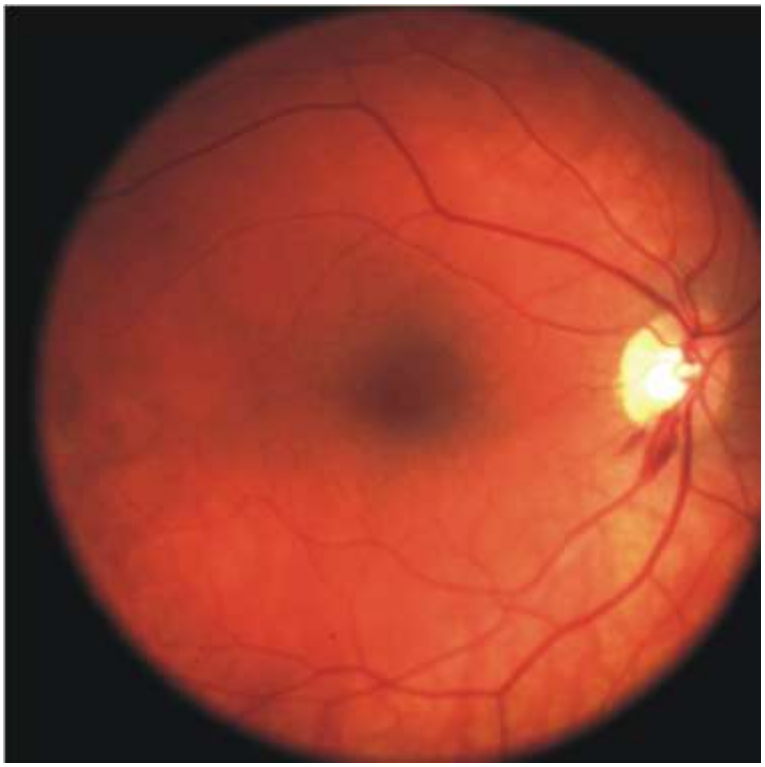
Slit Lamp
VH 4+, Concave Configuration Deep. Krukenburgs Spindle noted R&L. Spoke-like iris trans-illumination defects.

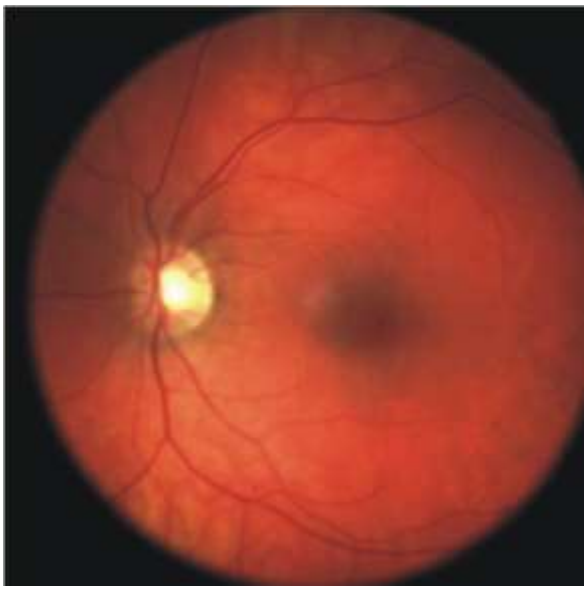
Gonioscopy : All quadrants wide open, iris configuration concave. TM Grade III – uniform with Sampolini Line

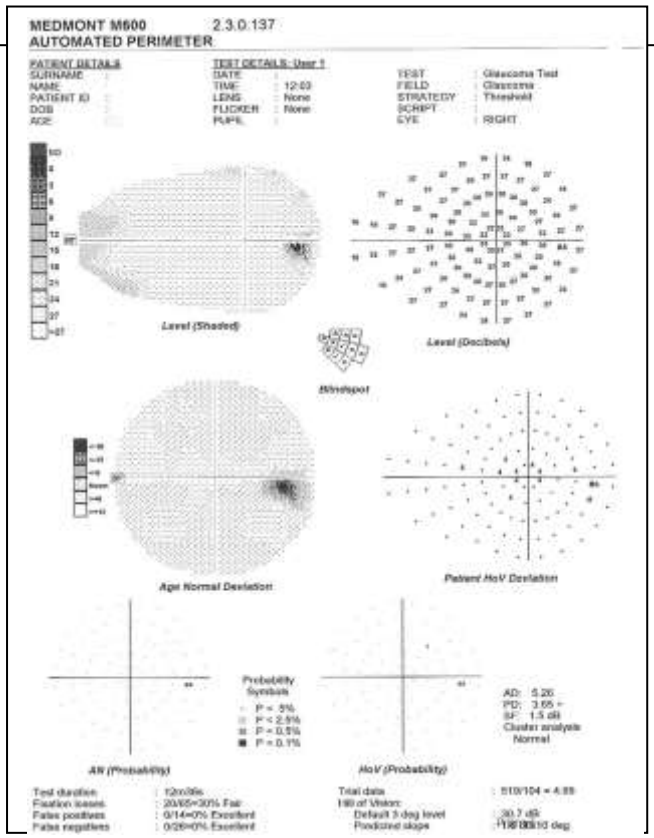
Dilated Fundsocopy (1% Tropicamide)
Nuclear Sclerosis. No Pseudo-exfoliation

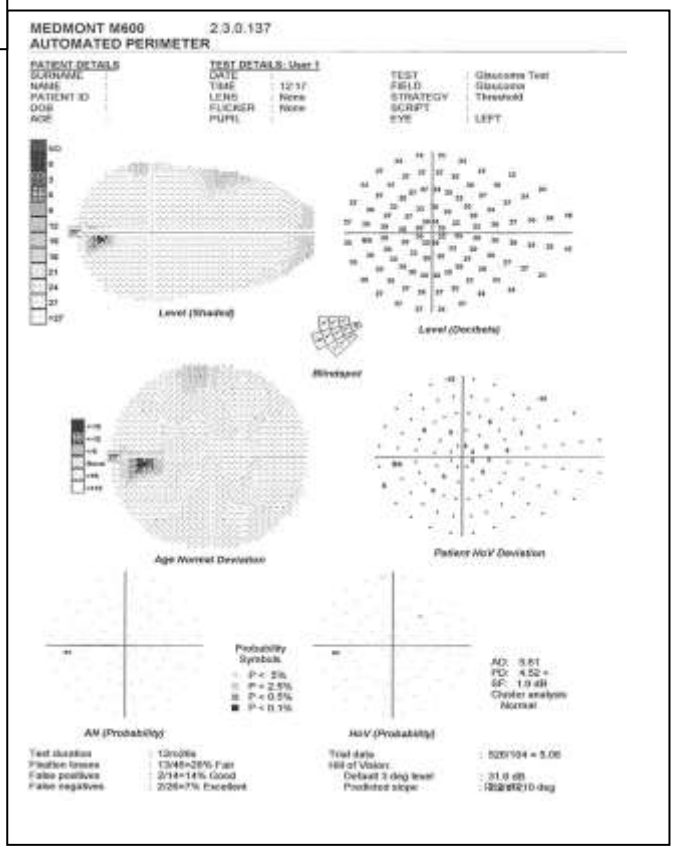
Right Disc - Splinter Haemorrhage inferior Rim. No inferior RNFL defect noted. VCD 0.5 Inferior rim obscured by Haemorrhage – no barring, no bayoneting, No PPA

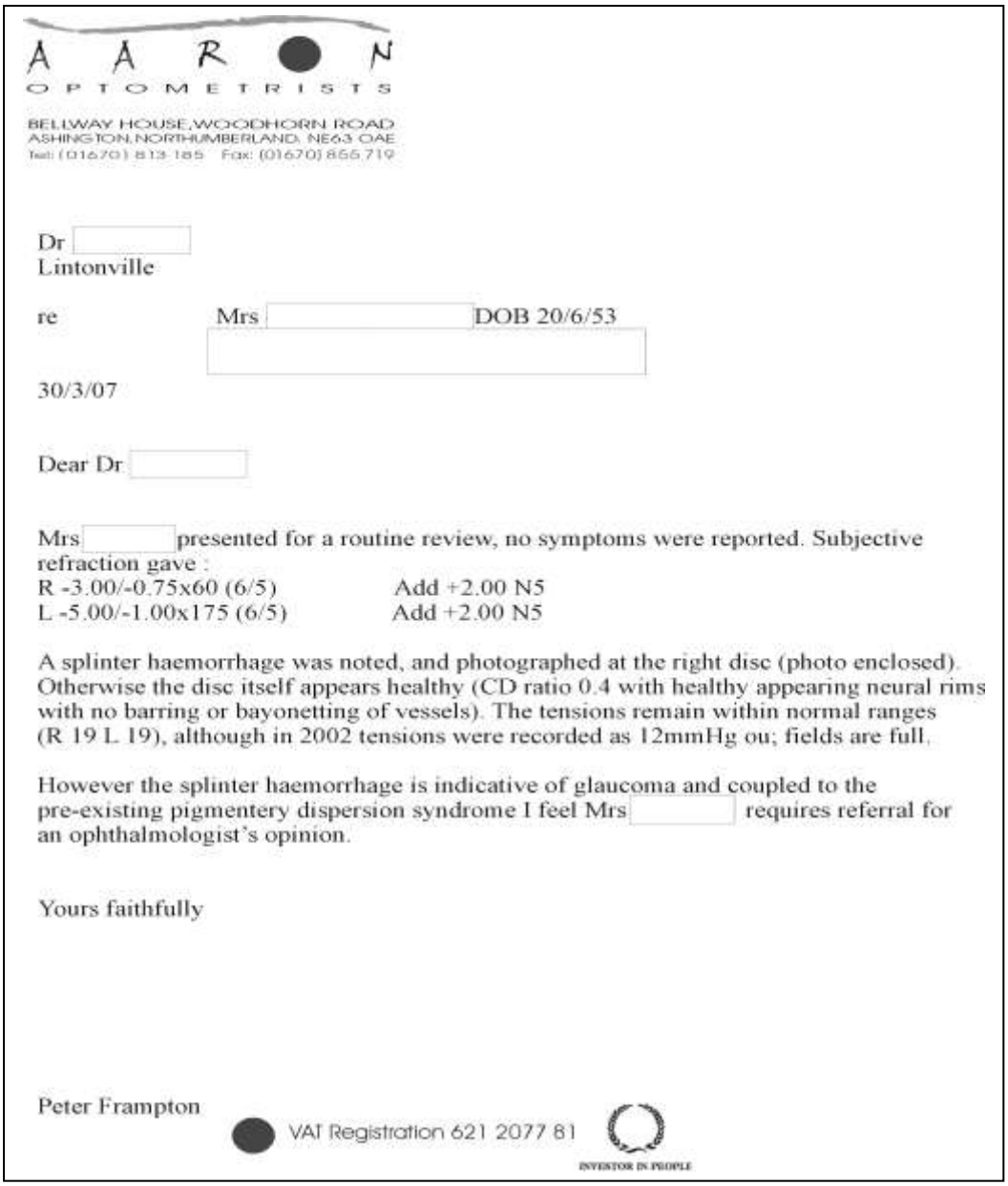
<u>Advice and CMP</u>
Routine referral to ophthalmology. PG/NTG/POAG?
Report sent to GP and copy given to Px. Outcome audit to confirm











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BELLWAY HOUSE,WOODHORN ROAD
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Tel: (01670) 813 185 Fax: (01670) 856 719

Dr
Lintonville

re Mrs DOB 20/6/53

30/3/07

Dear Dr

Mrs presented for a routine review, no symptoms were reported. Subjective refraction gave :
R -3.00/-0.75x60 (6/5) Add +2.00 N5
L -5.00/-1.00x175 (6/5) Add +2.00 N5

A splinter haemorrhage was noted, and photographed at the right disc (photo enclosed). Otherwise the disc itself appears healthy (CD ratio 0.4 with healthy appearing neural rims with no barring or bayonetting of vessels). The tensions remain within normal ranges (R 19 L 19), although in 2002 tensions were recorded as 12mmHg ou; fields are full.

However the splinter haemorrhage is indicative of glaucoma and coupled to the pre-existing pigmentery dispersion syndrome I feel Mrs requires referral for an ophthalmologist's opinion.

Yours faithfully

Peter Frampton

VAT Registration 621 2077 81

INVESTOR IN PEOPLE

At this review tensions were still within normal limits, although significantly higher than originally recorded in 2002. Fields were still excellent but a splinter haemorrhage was noted at the right disc.
The patient was referred for ophthalmologic assessment.

A splinter haemorrhage is highly suggestive of glaucoma (Werner 1996). The European Glaucoma Society further indicates that these haemorrhages are more common with Normal Tension Glaucoma (NTG), although this was simply drawing a comparison with Primary Open Angle Glaucoma (POAG) without consideration of PG.
With tensions within the normal range, should a differential diagnosis consider the possibility of NTG manifesting in a patient with PDS, or did the increasing pressure reflect a gradually congesting TM?

Campbell and Schertzer (1996) suggest the Trabecular Meshwork (TM) is dramatically altered in PG. In PDS the endothelial cells of the trabecular beams phagocytose the particulate load to excellent effect with preservation of the intertrabecular spaces and outflow facility.
The endothelial cells appear to be lifted loose in PG and there is associated trabecular collapse and loss of outflow facility. While reduced outflow facility is implicated in most glaucomas (Toris and Camras 2007), Werner (1996) implies it is near normal in NTG. The fundamental difference in pathophysiology could impact on management strategies.

Since outflow facility is rarely measured (Dueker 1996) other observations need to be relied upon. A more accurate assessment of PDS activity would be highly recommended. Amongst the signs of PDSa is 1) increasing number of transillumination defects, 2) increasing density of the Krukenberg Spindle, 3) increasing pigment on the anterior iris surface, 4) increasing trabecular meshwork pigmentation, 5) increasing IOP in association with reduced aqueous outflow facility (Campbell and Schertzer 1996).

No attempt at grading Krukenberg Spindle density, extremely difficult without a standardised grading scale, was made. Even with a standardised illumination technique, quantification of the number of iris defects would be confounded by iris pigment levels and pupil responses. With no clinical technique to estimate outflow facility, grading of TM pigmentation levels should have been attempted as the only practical way of grading progression. A pigment scale gonioscopy lens is available (Campbell and Schertzer 1996) but is certainly not commonly available in Optometric practice.
Using the gonioscopic grading scale described by Campbell and Schertzer (1996), the TM pigmentation was recorded as 3 (Dark Brown). However, this is a coarse scale and without a baseline is of little value. An attempt at grading the TM should have been made on 2002.

Other provocative tests, in particular, exercise (Campbell and Schertzer 1996, Ritch and Liebmann 1996), pupil dilation (Ritch 1982) and blinking (European Glaucoma Society 2003), can elicit significant elevations in IOP in some PG patients. A negative response, however, cannot rule out PG.

Ophthalmology post referral

Since referral the patient has been treated as a PG/NTG suspect and has been prescribed Xalatan® nocte.

Ritch and Liebmann (1996) suggest that treatment should be aimed at reducing pigment shedding as much as reducing IOP. The general approach to treatment is medical treatment first, laser second, and, finally, incisional surgery (Campbell and Schertzer 1996 European Glaucoma Society 2003).

Ritch and Liebmann (1996) suggest that by eliminating the iris concavity and iris/zonule contact, miotic therapy may prevent progression of the disease and even allow previously existing damage to reverse. The authors acknowledge that a serious caveat to this option is that most PDS patients are young and cannot tolerate pilocarpine drops.

Prostaglandins increase uveoscleral outflow (Camras 1996); while not reducing pigment shedding this would seem the logical first line medical treatment when conventional outflow is congested.
The only contraindication for Xalatan® listed in the Summary of Product Characteristics (SPC) of the Electonic Medicines Compendium (www.emc.medicines.org) is a 'known hypersensitivity to any component in Xalaltan®'. Further, prostaglandins require single daily administration (Phelan 2002) so aiding compliance (Watson 1998), and show very few side effects (BNF 2011, Camras 1996).

After three years of treatment with Xalatan® nocte, pressures of 14mmHg have been maintained.

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APPENDIX 1

Referral criteria for use by Optometrists

- IOP alone : (i.e. Normal fields and disc appearance)
  - 30 mm of Hg
  - 25 mm with family history

- Visual Fields (VF):

Normal: If disc suspicious or cupped and high IOP
Abnormal: If suspicious disc and/or high IOP
If normal disc and IOP : repeat field : only refer if repeatable

- Disc Appearance:
  - Pathological cupping
  - Disc haemorrhage
  - Disc Asymmetry only with high IOP and/ or Visual Field loss

- High IOP and anterior segment signs of secondary glaucoma eg pseudoexfoliation

- Suspect narrow angle glaucoma
  - Subacute attacks
  - Closeable angle and high IOP

Who can be discharged back to monitoring by their optometrist?

- Ocular hypertension with IOP < 30mm Hg or < 25mmHg with family history
- Untreated normal tension glaucoma with IOP < 18mm Hg in elderly patients.

Annual follow up by optometrist with VF. Follow up without VF acceptable if unable to do reliable fields in elderly with NTG.