Fuchs’ Uveitis and Posner-Schlossman Syndrome

A. Fuch’s Uveitis

Carl P Herbort, Nadia Bouchenaki

INTRODUCTION AND HISTORICAL ASPECTS

In 1906 Ernst Fuchs described a condition including heterochromia, unilateral inflammatory signs such as fine “microgranulomatous” keratic precipitates (KPs) as well as unilateral degenerative signs including iris atrophy and vitreous strands. He called the disease heterochromic cyclitis, more precisely his words were “cyclitis associated with heterochromia”, which would later be known by the eponym of Fuchs’ heterochromic cyclitis or iridocyclitis (Figures 1A and B).

In the 15th edition of Fuchs’ ophthalmology textbook the full clinical picture is described including the very prominent vitreous involvement and infiltration (Figure 2).

Because the disease was described in Vienna, in a Caucasian population, the most striking clinical sign, heterochromia, was included in the eponym of the disease called until today Fuchs’ heterochromic cyclitis. It is thought that in the past the disease was probably underdiagnosed in areas where irises are nearly exclusively brown and heterochromia is not part of the disease picture. The disease should therefore simply be named “Fuchs’ uveitis”. The disease pattern with onset mostly from the second to the fifth decade, points towards an external, probably infectious trigger that is at the origin of the subsequently autoentertained disease in persons that are probably genetically predisposed.3

EPIDEMIOLOGICAL ASPECTS

The disease seems to be occurring in most parts of the world with similar frequencies. In European reports, the percentage of cases in uveitis series is going from 2.2% in Portugal,4 5.0% in the Netherlands,5 5.4% in Switzerland6 to even 8.32% in Italy.7 Around the world, the proportion of Fuchs’uveitis (FU) in uveitis patient series is 6.6% in Iran,8 3.5% in Saudi Arabia,9 2% in Japan,10 5.7% in a recent study performed in southern China11 and 4.2% in African American patients.12 As can be concluded from this sample of epidemiological data, the disease seems to be ubiquitous and seems nowadays to be diagnosed with equal performance in brown iris population countries as shown by the recent chinese series.11

CLINICAL SYMPTOMS AND SIGNS

In contradiction to what is written in many textbooks and articles, Fuchs’ uveitis is a granulomatous disease. It has several clinical features such as Koepppe nodules and granulomatous keratic precipitates (KPs) that make up the definition of granulomatous uveitis. It should be remembered that the term granulomatous is a misnomer as terminology coming from pathology is used to describe a clinical entity or group of diseases. This term however serves very well clinical purposes as the definition of granulomatous uveitis is very precise and useful, including KPs that are more than
just dust on the endothelium, Koepp nodules at the pupillary margin and Busacca nodules on or within the iris. It is worthwhile to clearly specify here that as soon as KPs can be individually identified, meaning that they have already an “architecture”, they should be termed as granulomatous, which is not always clearly stated in textbooks. This is at the origin of the erroneous misclassification of Fuchs’ uveitis as a non granulomatous uveitis. In Fuchs’ uveitis the granulo-

Figure 2: Original text on Cyclitis associated with heterochromia in the 15th edition of Ernst Fuchs’ ophthalmology textbook

matous KPs have usually a quite specific aspect with fluffy edges taking sometimes a stellate appearance with fine spicules extending from the body of the KP
with a diffuse distribution on the endothelial surface (Figure 3).

**SYMPTOMS**

Most often floaters as well as blurred vision and visual deterioration is pushing patients to consult. Very often however the unilateral inflammation is a chance discovery during a routine ophthalmological examination. If anterior involvement is not prominent it can be missed and the attention of the ophthalmologist is attracted by the more obvious vitreal involvement often causing the diagnosis to be missed in favour of intermediate uveitis or other diagnoses such as the non specific diagnosis of “granulomatous uveitis”, “posterior uveitis” or “panuveitis”.

**SIGNS (TABLE 1)**

The signs that have mostly been put forward in the literature is heterochromia because it is the most spectacular sign as well as stellate granulomatous KPs that are also very characteristic. This is at the origin of the idea in the practitioner’s mind that Fuch’s uveitis is a purely anterior uveitis. If series coming from European countries are analysed, heterochromia is reported in 82% in the Netherlands,13 in 87% in a European study,14 in 80% in a North-American study15 and 70% in a United Kingdom study.16 When analysing studies coming from predominately brown eyed population countries the proportion of heterochromia is by far lower and goes down to 14% in China17 and to 23% in Japan.10 Instead of focussing on heterochromia, the iris signs to search for are iris atrophic changes with or without heterochromia. In some series atrophic iris changes are present in close to 100% of cases (Figure 3).

The proportion of patients for whom characteristic KPs were described do not differ among Caucasian and brown eyed populations representing respectively 88% (Netherlands), 13 96% (Belgium), 14 81% (UK)16 and 96% in a North American study15 which is not sensibly different for Asian series where the percentage is respectively 91.5% for China17 and 97% for Japan10 (Figures 4A-C). See proportion of signs in table one.18-20

Another set of signs is seen less consistently as the two previous signs but is very helpful to make the diagnosis more certain. This includes posterior polar subcapsular cataract (Figure 5), iris (Koeppe > Bussaca) nodules (Figure 6) and abnormal vessels in the irido-corneal angle. The latter sign is not always reported in Fuchs’ uveitis series. These abnormal angle vessels are also at the origin of the classically reported “Amsler’s sign” consisting of occurrence of a hyphema when an anterior chamber puncture or tap is performed. Similarly these vessels are also at the origin of hyphema during cataract surgery.

One of the most neglected signs is vitreous infiltration which is even not reported in some series, although it is one of the signs described originally by Fuchs (Figure 7).

The degree of vitreous involvement may vary from the presence of a few cells to an infiltration with dense vitreous fibrous strands and condensations which may explain an erroneous diagnosis such as intermediate or posterior uveitis. In rare cases the heavily infiltrated vitreous may be at the origin of peripheral retinal tears and retinal detachment.

Finally absence of some clinical signs are almost diagnostic and may be excluding signs if present but are rarely reported. This is the case of “absence of cystoid macular oedema” (CMO) and “absence of posterior synechiae” on the crystalline lens which are present if reported in a proportion of over 95%. In the
<table>
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<tr>
<th>Country</th>
<th>Switzerland</th>
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<th>Belgium</th>
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<tr>
<td>Name 1st author</td>
<td>Bouchenaki</td>
<td>La Hey</td>
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<td>Nbr of Cases</td>
<td>88</td>
<td>51</td>
<td>550</td>
<td>77</td>
<td>103</td>
<td>26</td>
<td>54</td>
<td>132</td>
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<td>FHC</td>
<td>FHC</td>
<td>FHC</td>
<td>FHU</td>
<td>FHC</td>
<td>FHC</td>
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<td>14</td>
<td>16</td>
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<td>20</td>
<td>15</td>
<td>18</td>
<td>10</td>
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<tr>
<td>Vitritis</td>
<td>98%</td>
<td>84%</td>
<td>69%</td>
<td>84%</td>
<td>66.60%</td>
<td>14.80%</td>
<td>55%</td>
<td>N/D</td>
<td>88%</td>
<td>73.8</td>
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<td>Fuchs KPs</td>
<td>85%</td>
<td>88%</td>
<td>96%</td>
<td>81%</td>
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<td>100%</td>
<td>96%</td>
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<td>43%</td>
<td>82%</td>
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<td>70%</td>
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<td>94%</td>
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<td>22%</td>
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<td>N/D</td>
<td>N/D</td>
<td>N/D</td>
<td>N/D</td>
<td>70%</td>
<td>N/D</td>
<td>33%</td>
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<td>32%</td>
<td>20%</td>
<td>7.40%</td>
<td>N/D</td>
<td>N/D</td>
<td>83%</td>
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<tr>
<td>Bilaterality</td>
<td>12%</td>
<td>4%</td>
<td>8%</td>
<td>15.80%</td>
<td>7.80%</td>
<td>4%</td>
<td>0%</td>
<td>21%</td>
<td>0%</td>
<td>13.50%</td>
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<tr>
<td>Absence of CMO</td>
<td>100%</td>
<td>98%</td>
<td>N/D</td>
<td>N/D</td>
<td>N/D</td>
<td>N/D</td>
<td>N/D</td>
<td>N/D</td>
<td>98%</td>
<td>N/D</td>
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<tr>
<td>Absence of synechiae</td>
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<td>100%</td>
<td>N/D</td>
<td>98.70%</td>
<td>100%</td>
<td>N/D</td>
<td>N/D</td>
<td>N/D</td>
<td>94%</td>
<td>81%</td>
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FHC = Fuchs' Heterochromic Cyclitis
FHU = Fuchs' Heterochromic Uveitis
N/D = not done
absence of the characteristic signs of heterochromia and Fuchs' KPs and the presence of vitritis these signs have almost determining diagnostic value. However both these signs are reliable only in patients that have never undergone intraocular surgery. Fuchs' uveitis patients having had intraocular surgery are no more protected against CMO nor irido-lenticular synechiae. The proportion of any given sign in Fuchs' uveitis series is shown on Table 1, indicating some diversity in the proportion of reported signs from one study to another.
Imaging in Uveitis

LASER FLARE PHOTOMETRY

Laser flare photometry (LFP) is measuring back scattered photons from anterior chamber proteins when a laser light beam is shone into the anterior chamber. Its principle is identical to slit-lamp anterior chamber examination except that in LFP the light source is monochromatic and quantified rather than polychromatic. Also the light detector is objective made of a photomultiplyer and photodetector rather than the subjective human eye. This method therefore allows to measure objectively the disruption of the blood-aqueous barrier and hence the exact level of intraocular inflammation.

When we measured flare in our Fuchs’ uveitis patients at presentation the mean flare value was 8.62 ± 4.3 photons per millisecond (ph/ms) which is only slightly above normal values (4-6 ph/ms). This showed that in most Fuchs’ cases there is minimal disruption of the blood-aqueous barrier, hence minimal anterior chamber inflammation. Moreover, at the end of follow-up this value remained stable at 8.45 ± 4.3 ph/ms in our patients and did not increase, showing that inflammation does not progress in the absence of treatment. This has recently been confirmed by a chinese study reporting similar values.

It is however in every day practice that LFP has proved especially useful for Fuchs’ uveitis. In those Fuchs cases that have been misdiagnosed and are under topical and sometimes even systemic corticosteroid therapy, LFP allowed to show that, after discontinuation of therapy, there is no rebound flare increase or minimal flare increase indicating to the patient and to the referring doctor that it is safe to withdraw potentially harmful therapy that has no impact on inflammation in this disease.

ANGIOGRAPHIC FINDINGS

In case of a clinically sure diagnosis of Fuchs’ uveitis there is no need to perform fluorescein angiography. Very often however diagnosis is missed initially and fluorescein angiography happens to be performed. We analysed a series of 23 patients that had a fluorescein angiography performed for diagnostic purposes.

In all but one of these patients was there a slight to moderate and sometimes pronounced disc hyperfluorescence (Figure 8). The only patient who had no disc hyperfluorescence was a case that was known to have disc atrophy previous to the diagnosis of Fuchs’ uveitis. In 4 patients slight mid-peripheral retinal vascular leakage was seen. No cystoid macular oedema could be seen in any of the 21 patients that had never had intraocular surgery even after very longstanding inflammation. Cystoid macular oedema was present in two patients both of whom had undergone cataract surgery.

Being aware of these results it might be helpful and justified to use fluorescein angiography as a diagnostic procedure in some cases where Fuchs’ uveitis is the presumed diagnosis but where some of the principal signs are missing. In such cases with longstanding uveitis with prominent vitreous infiltration, the absence of an angiographic CMO together with disc hyperfluorescence can be considered confirmatory of Fuchs’ uveitis.

DIFFERENTIAL DIAGNOSIS, EVOLUTION, THERAPY AND PROGNOSIS

In the paragraphs on differential diagnoses given for Fuchs’ uveitis, textbooks usually only cite other anterior uveitis entities such as herpetic uveitis, zoster uveitis, Posner Schlossman syndrome, etc. Only one
textbook cited intermediate uveitis as a possible differential diagnosis. This shows how strong the idea is, even among textbook authors, that Fuchs’ uveitis is a purely anterior uveitis, making abstraction of the vitreous involvement. In real ophthalmologic practice the situation is quite different. In our series in which 84% of patients were referred, the list of erroneous diagnoses, hence diagnoses that should be listed in the differential diagnosis for Fuchs’uveitis, was headed by intermediate uveitis in 37/59 (63%) of non diagnosed cases followed by panuveitis in 11/59 (19%), posterior uveitis in 6/59 (10%) patients and granulomatous uveitis in 5/59 (8%). The nature of these diagnoses indicates that the clinician tends to rule out Fuchs’ uveitis in the presence of inflammatory vitreous involvement.

Evolution of Fuchs’ uveitis is mostly benign as the sensitive structures such as the macula is nearly never involved despite substantial and prolonged vitritis. Therefore no therapy is necessary. Nevertheless regular follow-up examinations are necessary in order to detect complications such as intraocular hypertension or glaucoma as well as cataract which may be part of the natural course of the disease but which are often favoured by the abusive and/or long term use of steroids. Prognosis is rather good because even if cataract is present, it is operated with success because these eyes tolerate surgery very well as was noted in Fuchs’ original descriptions. Among the 10 to 20% of patients that develop glaucoma, some are difficult to stabilise despite surgery and prognosis is reserved only in this portion of Fuchs patients.

Despite the fact that about 15-20% of Fuchs’ eyes develop hypertension, the mean pressure of the Fuchs eye is lower than the fellow eye (12.04 ± 4 versus 12.87 ± 4.2 mmHg), indicating that, as a rule, the affected eye has a lower pressure than the normal eye.

Although corticosteroid therapy is not supposed to be used because of absence of impact, there are rare situations where inflammatory bouts occur that might benefit from short courses of topical steroids.

WHY IS FUCHS’ UVEITIS SO OFTEN MISDIAGNOSED OR DIAGNOSED WITH SUBSTANTIAL DELAY? / DIAGNOSTIC CRITERIA OF FUCHS’ UVEITIS

The reasons why Fuchs’ uveitis is so often misdiagnosed or underdiagnosed are several, including bilaterality usually not associated with the disease but which occurs in about 10%, search for heterochromia which is often absent especially in brown eyed countries and absence of the characteristic KPs. The main reason however, as stated earlier, is the presence of inflammatory vitreous involvement that, in the mind of the practitioner, is usually not associated with Fuchs’ uveitis when in reality it is a major diagnostic sign. This leads to diagnostic delay which in our series occurred in 59/79 (75%) cases with a mean duration of delay of 3.67 ± 4.34 years. Unawareness that vitritis is
part of Fuchs’ clinical picture is also at the origin of inadequate, useless and potentially harmful therapy such as systemic corticosteroids and/or even systemic immunosuppressants that were given in 34/79 (43%) patients in our study.

Therefore diagnostic criteria of Fuchs’ uveitis have to be reconsidered. The most striking clinical signs such as heterochromia and characteristic KPs are of course very useful for the diagnosis when they are present. However the diagnosis has to be made more secure by considering also less spectacular signs albeit highly consistent that put together allow to make the diagnosis of the condition even without the more obvious signs. A very high, almost 100% degree of certainty is obtained when triad consisting of heterochromia, typical KPs and vitritis is present. The neglected or more discrete signs that are recorded with a proportion of close to 100% in Fuchs’ series are, when going antero-posteriorly (1) atrophic iris changes, (2) absence of posterior irido-crystalline synechiae, (3) vitritis or vitreous strands and (4) absence of cystoid macular edema. The occurrence of the combination of these signs probably allows to reach an almost identical degree of certainty. The association of “minor signs” such as (1) iris nodules, (2) cataract and (3) stromal pseudo-neovessels apparent because of iris atrophy or abnormal iridocorneal angle vessels with or without Amsler’s sign (hyphema following anterior paracentesis) add to the degree of certainty of the diagnosis.

**PATHOGENESIS OF FUCHS’ UVEITIS AND CONCLUSION**

Fuchs’ uveitis typically has the profile of a disease caused by an infectious triggering factor (probably more than one) at the origin of an immune auto-entertained disease in genetically susceptible patients. Most cases are diagnosed between the 2nd and the 5th decade, the period during which individuals are exposed to most of the infectious agents encounter throughout life including the agent(s) that can trigger Fuchs’ uveitis.13,15,19 Above the age of sixty the disease is less frequently diagnosed as the probability to encounter the infectious trigger is much less. HLA-B27 related acute anterior uveitis has an identical epidemiological profile. For this disease the triggering agents are known to be debris from gram-negative bacteria and the genetical predisposition is the presence of the HLA-B27 antigen.

The evidence accumulated so far indicating immune dysfunction in Fuchs’ patients and Fuchs’ eyes is sufficient to allow us to consider immune mechanisms as the common effector pathway causing lesions in Fuchs’ uveitis. Very early, when immunological investigations were still very rough, increased IgG in aqueous humour was found.26 This was later confirmed by PI Murray et al who also found increased interleukin-6 in the aqueous humour.27,28 Other studies showed the presence of autoimmunity to corneal antigens, a possible explanation of the spread-out disposition of Fuchs KPs. Immunohistochemical analysis of iris biopsy specimens from patients with Fuchs’ heterochromic cyclitis showed evidence of an inflammatory cell infiltrate, which was a mixture of interleukin-2 receptor-negative T helper and suppressor cells, B lymphocytes, and plasma cells. More recently Labalette and colleagues showed the presence of CD8-positive CD28-negative T cell clonotypes indicating an antigen-driven process involving these cells at the origin of Fuchs’ uveitis.31 In addition a cellular autoimmune response to S antigen was found in patients with Fuchs’ uveitis, a possible explanation of posterior segment involvement,32 which seems to have received clinical support by a study that showed subclinical retinal damage using electroretinography.33

As far as the infectious trigger in Fuchs’ uveitis is concerned, several agents have been put forward. For some time toxoplasma Gondii had the favours.34,35 only to be seriously put in doubt some years later.36 Recently the detection of an intraocular synthesis of anti-rubella antibodies in all tested patients as well as the rubella genoma itself in 20% of eyes, represented a very strong argument for Fuchs’ uveitis being a rubella virus driven uveitis.37 Similar results were also found in another group.38 Although there are presently quite convincing arguments for rubella to be a trigger for Fuchs’ uveitis, other possible triggers might also play a role.

In conclusion, Fuchs’ uveitis is an ubiquitous disease involving the vitreous and the anterior uvea. The inflammatory nature does make no doubt and the presently favoured pathogenetic mechanism is an immune process triggered by an infectious agent thought to be in many cases rubella virus, other agents
not being completely excluded. The disease is often misdiagnosed because of the textbook generated unawareness of the quasi 100% rate of association of vitritis with the disease. Anti-inflammatory treatment has mostly no or minimal impact but evolution is favourable unless complications such as glaucoma or cataract occur.

**KEY POINTS**

- Fuchs’ uveitis is a mostly unilateral uveitis involving the anterior segment and the vitreous body.
- It is usually diagnosed between the second and fifth decade, a disease pattern indicating the association of an external trigger associated with susceptibility to the disease.
- Moderate decrease of visual function and floaters are the main symptoms but very often the disease is diagnosed by chance during a routine visit.
- The main signs of Fuchs’ uveitis with a proportion of presence between 90-100% are (1) characteristic Fuchs’ KP, (2) iris atrophic changes, (3) vitritis, (4) absence of synechiae, (5) absence of CMO, (6) unilaterality
- Complications include cataract formation (> 60%, depending on length of follow-up) and glaucoma (12-20%)
- No treatment is necessary for the primary disease.

**REFERENCES**


INTRODUCTION

Posner Schlossman syndrome (PSS) as originally described in 1948 by Posner and Schlossman is a self-limiting and benign condition characterized by unilateral, recurrent attacks of mild, granulomatous iritis with elevated intraocular pressure (IOP) during the acute attack, open angle, normal visual field and optic disc.\(^1\)

It is classified as an inflammatory glaucoma because it is by definition always accompanied by uveitis.\(^1\) The inflammation may follow the acute rise in intraocular pressure (IOP) by some days or be so mild as to be overlooked, so the etiology of the rise in pressure may not be clear at first presentation.

EPIDEMIOLOGY AND PATHOGENESIS

PSS tends to affect patients between 20 and 50 years of age. Males are more often affected. There is no reported ethnic or racial predilection. Recent reports suggest an association with the human leukocyte antigen (HLA) Bw54.\(^2,3\) Recurrence is common and can be in either eye but intervals between attacks tend to increase during the course of the disease.\(^4\)

The etiology of PSS is uncertain and no recognized associated systemic conditions have been reported. The mechanism responsible for glaucomatocyclitic crisis is unclear. Cytomegalovirus (CMV), varicella-zoster virus and herpes simplex virus (HSV) may be linked to the disease.\(^5\) The trabecular meshwork is innervated by the trigeminal nerve and is a conduit for HSV. One postulated mechanism for the syndrome is that HSV causes an inflammation of the trabecular meshwork, impeding aqueous outflow and increasing IOP.

The allergic response is also under investigation. High levels of prostaglandin E have been found in the aqueous humor during attacks. These levels have been observed to return to normal during remissions.\(^2,6\)

CLINICAL FEATURES

Typically, patients describe a history of intermittent episodes of mild visual blurring, colored haloes around lights, and minimal discomfort in one eye. Some patients have no symptoms.

The hallmark feature of PSS is that the IOP (often 45 mmHg or higher) does not correspond to the amount of ocular inflammation. The affected eye typically appears white and quiet on gross inspection, but a small amount of inflammation can usually be detected. The anterior chamber is deep. A few, small, round non-pigmented keratic precipitates (KPs) tend to accumulate on the lower one third of the corneal endothelium within three days of onset (Figure 1).
A common finding is that the pupil is slightly dilated in the affected eye compared to the unaffected eye. Gonioscopy reveals normal, open angles without abnormal pigmentation. Posterior and anterior synechiae do not develop in this condition. Signs may be present for a month or longer without producing substantial symptomatology.

PSS is a self-limiting condition that lasts a few weeks to a month. During the acute and remission phases of PSS, anterior chamber angles are typically open with normal pigmentary density and no associated synechiae. Although attacks are self limited, glaucomatous cupping and field loss may occur.

Recurrent attacks characterize the illness; however the frequency of such attacks is highly variable between patients. Usually only one eye is affected; rarely however, a patient may experience subsequent episodes in either eye.

Posner-Schlossman syndrome does not always follow a completely uncomplicated course.

Repeated episodes of elevated intraocular pressure can cause long-term sequelae such as glaucoma. In a study of 53 cases of PSS, 14 (26.4%) were found to have glaucomatous optic nerve damage.

**DIAGNOSIS**

There are no specific laboratory tests for this entity and a presumptive diagnosis of PSS was made based on the following findings: recurrent episodes of mild iritis associated with elevated IOPs, diffuse epithelial edema of the cornea, and a few fine keratic precipitates. The IOP is normal between attacks and the angles are open.

**DIFFERENTIAL DIAGNOSIS**

When diagnosing PSS, we must rule out other conditions that could cause unilateral or bilateral uveitis. These include Fuchs’ uveitis, acute angle-closure glaucoma, pigmentary glaucoma, herpes simplex virus infection, uveitic glaucoma, neovascular glaucoma, and secondary angle-closure glaucoma due to peripheral anterior synechiae.

History, biomicroscopy, tonometry and dilated fundoscopy are essential for the differential diagnosis. Past ocular, systemic, social and family histories are necessary to rule out suspected underlying etiologies.

**TREATMENT**

The treatment for PSS focuses on lowering IOP and reducing inflammation. Therapy may include topical beta blockers, topical alpha agonists topical/oral carbonic anhydrase inhibitors and even hyperosmotic agents. Myotic agents are contra-indicated for the treatment of PSS and uveitic glaucoma because they enhance the risk for the formation of posterior synechia, cause discomfort to the patient by aggravating ciliary muscle spasm and increase inflammation in the eye by amplifying the breakdown of the blood aqueous barrier. Pain and discomfort may be addressed with cycloplegic agents such as atropine 1%, homatropine 5%, or scopolamine 0.25%. Inflammation can be treated with topical steroidal preparations, every two hours depending on the severity of the episode. Prophylactic anti-inflammatory therapy is somewhat controversial. Most practitioners do not feel that it prevents recurrences.

**KEY POINTS**

- PSS is a condition characterized by unilateral attack of mild, granulomatous iritis with elevated IOP during the acute attack, white eye, open angle, normal visual field and optic disc.
- The etiology of PSS is uncertain, but several viruses, including CMV, VZV, and HSV have been suggested as potential causes of the syndrome.
Recurrent attacks characterize the illness. PSS is a self-limiting condition, but glaucomatous optic nerve damage may occur as consequence of repeated episodes of elevated IOP.

The treatment for acute attacks of PSS focuses on lowering IOP and reducing inflammation. Prophylactic anti-inflammatory therapy does not seem to prevent recurrences.

REFERENCES