Punctate Inner Choroidopathy (PIC) is a rare form of posterior uveitis. PIC is one of the so called ‘white dot syndromes’ which come under the heading posterior uveitis. PIC has only been recognised as a distinct condition, or type of posterior uveitis, as recently as 1984 when Watzke identified 10 patients who appeared to make up a distinct group within ‘white dot syndromes’.

Main features of PIC

- Typically affects short sighted (myopic) women. (90% of cases are female).
- The average age of patients with PIC is 27 years with a range of 16-40 years.
- Patients are otherwise healthy and there is usually no illness, which triggers the condition or precedes it.
- The inflammation is confined to the back of the eye (posterior). There is no inflammation in the front of the eye (anterior chamber) or vitreous (the clear jelly inside the eye). This is an important distinguishing feature of PIC.
- It usually affects both eyes.
- The appearance of gray-white or yellow punctate (punched out) areas (lesions) at the level of the inner choroid. These lesions are typically located centrally at the back of the eye (posterior pole).

Symptoms typically include:

1. Blurring of vision,
2. Partial ‘blind spots’ or scotoma. These areas of diminished or lost areas of the visual field are typically near the centre of vision but occasionally can be peripheral. These may be temporary or permanent.
3. Seeing flashing lights. This is known as photopsia.
Causes

There are no known causes of PIC, but may represent an autoimmune type of uveitis.

Diagnosis

Diagnosis of PIC can be difficult because the appearance may be similar to other conditions and types of posterior uveitis, especially other forms of the so-called ‘white dot syndromes’. The diagnosis is made by eliminating all the other possibilities by careful examination by an experienced uveitis specialist, aided with visual field testing and angiography, an intra venous dye used to show the blood vessels at the back of the eye (e.g., fluorescein or indocyanine angiogram).

It is important that the correct diagnosis is made because treatment may be quite different for apparently similar conditions. An example of this would be PIC and multifocal choroiditis and panuveitis (MCP). The 2 conditions share similar features (myopic women, shape of lesions etc.) but MCP has anterior and vitreal inflammation and so requires more aggressive treatment.

Course of the condition or ‘What usually happens’

What happens with PIC depends a lot on the presence or not of an important complication, choroidal neovascularisation (known as CNV). Often, the inflammation in PIC is self-limiting, not always requiring treatment. However, treatment is advised if there are many active or central lesions, or if there are signs of CNV.

What is CNV?

The PIC lesions, which form scars deep in the choroid layer of the eye, may result in new blood vessels forming. These can be seen as the body’s attempts at repair, but these new blood vessels (neovascularisation) are weak, can spread to form a membrane and can threaten the vision. It is suspected that at least 40% of patients with PIC develop CNV. This is a complication, which can occur in other white dot syndromes and other eye conditions such as macular degeneration but occurs rarely in other forms of uveitis.

CNV is a sight threatening complication and so must be picked up early and always treated. It may occur whether the uveitis is active or not. CNV, if not treated, may lead to subretinal fibrosis (scarring), a further complication, which is more difficult to treat, and which leads to poor vision.

Good monitoring for patients with PIC is therefore very important.
Treatment

It is important to distinguish between treatment of the underlying inflammation (PIC) and the treatment of CNV.

2-pronged approach

Treatment is not always necessary and observation may be appropriate for lesions if they are found in non-sight threatening areas (that is not centrally). Active lesions of PIC can be treated with corticosteroids taken systemically (tablets) or regionally by injections around the eye (periorbital). It has been argued that treating lesions in this way may help minimise the development of CNV.

The treatment of CNV

Early treatment is required for this complication. There are several possible treatment methods, but none of these treatments appears to be singly effective for the treatment of CNV.

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<tr>
<td>1</td>
<td>Corticosteroids</td>
<td>Regional or systemic or intraocular</td>
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<td>2</td>
<td>'Second line’ immunosuppressants</td>
<td>There is evidence that combined therapies of steroids and second line immunosuppressants may be important</td>
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<td>3</td>
<td>Surgical excision of the affected area</td>
<td>In well selected cases.</td>
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<td>4</td>
<td>Intravitreal anti-vascular endothelial growth factor (VEGF) agents.</td>
<td>Examples are bevacizumab (avastin) and ranibizumab (Lucentis). These relatively new drugs are injected into the eye</td>
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<td>5</td>
<td>Photodynamic therapy (PDT)</td>
<td>A photosensitive drug is 'activated' by strong light. Consideration may be given to combined therapy of PDT and anti VEGF.</td>
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<td>6</td>
<td>Laser photocoagulation</td>
<td>This is occasionally used unless the CNV is subfoveal (affecting the central or macular part of the vision). The laser treatment can damage the vision in this.</td>
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The use of the intravitreal anti VEGF agents namely bevacizumab (avastin) and ranibizumab (Lucentis) have been described recently. The current evidence supporting the use of anti VEGF agents is based on retrospective case studies and could not be described as strong. However, further data from prospective controlled trials are needed before the therapeutic role of anti-VEGF therapy in the uveitis treatment regimen can be fully determined. The anti VEGF agents furthermore have not been shown to have an anti-inflammatory effect.

Thus, treatment of the underlying inflammatory disease should play a central role in the management of uveitic CNV. A two-pronged treatment that focuses on achieving control of inflammation through the use of corticosteroids and/or immunosuppressive agents, while treating complications that arise despite adequate disease control with intravitreal anti-VEGF agents, may be useful.

Regular monitoring is essential to achieve a good outcome. This must be stressed. This is because even if there is no active inflammation, there may still be CNV which you may not be aware of but which requires treatment to avoid suffering vision loss.

Outcome / Prognosis

The visual prognosis of eyes with PIC that do not develop subfoveal CNV is good. If CNV is picked up early and treated appropriately then the visual outcome can also be good. Frequent monitoring is important to ensure a good outcome. Poor vision occurs mostly with subfoveal CNV or if subretinal fibrosis (scarring) has formed.

Further information

If you have been diagnosed with Punctate Inner Choroidopathy and want to contact another patient who has personal experience of the eye condition, Dave Stead of The PIC Society can be contacted via the PIC society discussion forum that he runs at: http://www.pic-world.net/

The information contained in this Factsheet has been written for patients by Phil Hibbert and Dave Stead (both patients) It has been checked by Professor Andrew Dick, of Bristol Eye Hospital, and UIG Professional panel chairman. Published by Uveitis Information Group (UIG) May 2011

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