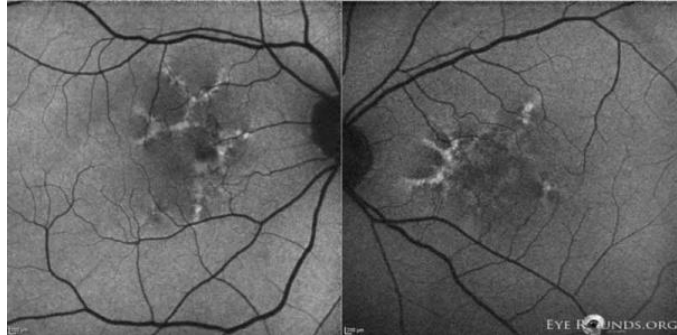


**Butterfly-shaped pattern dystrophy** is a rare **inherited retinal disorder** that falls under the category of **pattern dystrophies of the retinal pigment epithelium (RPE)**. This condition is characterised by a **butterfly-shaped pigmentation pattern** in the macula, visible during fundus examination.

## Key Features:

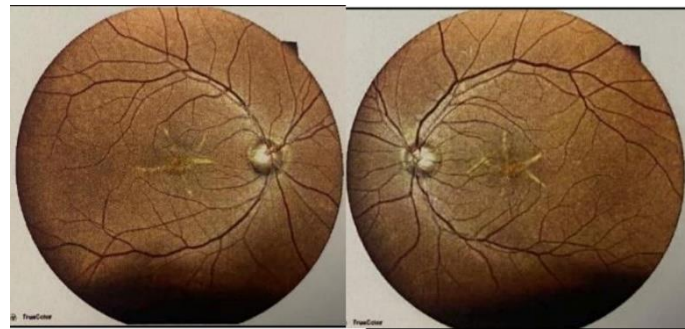
### 1. Appearance:

- Pigment deposits in the macula form a **bilateral butterfly or bowtie-like shape**.
- These deposits are due to the abnormal accumulation of **lipofuscin** or other metabolic by-products in the retinal pigment epithelium.



### 2. Cause:

- Typically caused by mutations in the **PRPH2 (peripherin-2)** gene, which plays a role in maintaining photoreceptor disc structure.
- Inherited in an **autosomal dominant pattern**, meaning one copy of the mutated gene can lead to the condition.



### 3. Symptoms:

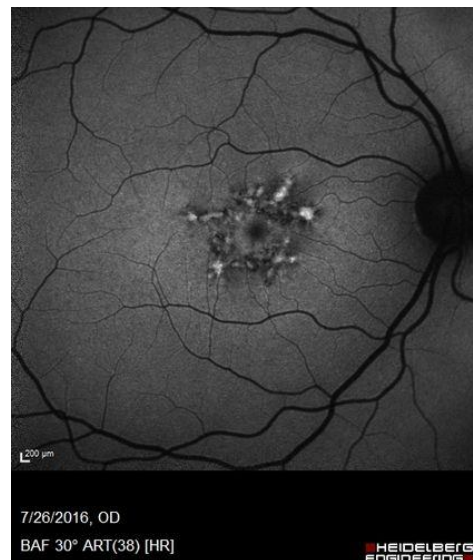
- Many patients remain **asymptomatic** in the early stages.
- Later stages may involve:
  - **Mild vision loss.**
  - **Metamorphopsia** (distorted vision).
  - Difficulty with night vision or central vision as the condition progresses.

### 4. Natural History:

- Progression is generally **slow**, and severe vision loss is uncommon.
- In some cases, **choroidal neovascularisation (CNV)** may develop, potentially leading to more significant vision changes.

### 5. Diagnosis:

- **Fundus examination:** Reveals butterfly-shaped pigment abnormalities in the macula.
- **Optical coherence tomography (OCT):** May show disruption of the outer retinal layers and RPE.
- **Fundus autofluorescence (FAF):** Highlights the areas of lipofuscin accumulation.
- **Genetic testing:** Confirms PRPH2 or related mutations.



### 6. Differential Diagnosis:

- Adult-onset vitelliform macular dystrophy.

- Age-related macular degeneration (AMD).
- Other pattern dystrophies.

#### 7. Management:

- No specific treatment is available for butterfly-shaped pattern dystrophy.
- **Monitoring:** Regular follow-up is needed to detect complications like CNV early.
- **Low vision aids** may help in advanced cases.
- In CNV cases, anti-VEGF injections (like in AMD) may be used.

#### Prognosis:

- The condition generally has a **good visual prognosis**, but regular monitoring is essential to manage potential complications.

**Table 1. Five Types of Autosomal Dominant Pattern Dystrophies and Typical Characteristics**

Dystrophy	Typical Age of Onset	Typical VA	Retinal Presentation
Adult-Onset Vitelliform Dystrophy	4th-6th decades	20/30-20/60	Bilateral, circular, 1/3-1DD
Butterfly-Shaped Pigment Dystrophy	2nd-5th decades	20/20-20/25	Bilateral, triradiate hyperpigmentation
Reticular Dystrophy	5th decade	20/30-20/70	Bilateral, fishnet/ chicken wire hyperpigmentation pattern, 4-5DD
Multifocal Pattern Dystrophy	4th-6th decades	20/20-20/40	Bilateral, multiple yellow fleck-like lesions
Fundus Pulverulentus	4th-5th decades	20/20-20/40	Bilateral, coarse macular pigment mottling

Good article : <https://www.reviewofoptometry.com/article/pattern-recognition-how-to-identify-and-confirm-multifocal-pattern-dystrophy>