



Plain Language Summary

Long-term Visual Outcomes in Pseudoxanthoma Elasticum-associated Choroidal Neovascularization: A National Database Study

Plain language summary by Asha Depoy, PXE International Volunteer

Study at a Glance

Long-term Visual Outcomes in Pseudoxanthoma Elasticum-associated Choroidal Neovascularization: A National Database Study

Lead Institution:

Retina Service, Wills Eye Hospital, Mid Atlantic Retina, Thomas Jefferson University, Philadelphia, Pennsylvania.

Collaborating Institution: Vestrum Health, Naperville, Illinois.

Study Type: Retrospective database study

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Citation: Zafar S, Aggarwal N, Winsor J, Sina E, Vemula S, Wibbelsman TD, Boucher N, Yonekawa Y. Long-term Visual Outcomes in Pseudoxanthoma Elasticum-associated Choroidal Neovascularization: A National Database Study. *Ophthalmic Surg Lasers Imaging Retina*. 2026 Feb;57(2):112-116. doi: 10.3928/23258160-20251022-01. Epub 2025 Dec 15. [PMID: 41401369](#).

Summary:

Why was this study done?

One of the most serious eye problems in pseudoxanthoma elasticum (PXE) is **choroidal neovascularization (CNV)**. CNV occurs when abnormal blood vessels grow beneath the retina. These vessels can leak or bleed, damaging central vision.

Today, CNV is usually treated with medications called **anti-VEGF injections**, which are injected into the eye. While these treatments have greatly improved outcomes, little has been known about how people with PXE do over many years in routine clinical practice.

This study examined how often people with PXE need treatment and how well their vision is preserved over time.

What did the researchers do?

The researchers analyzed information from a large U.S. retina database that included patients seen between **2015 and 2024**.

They studied:

- **171 people with PXE**
- **248 affected eyes**
- Average follow-up of about **3 years**

They recorded:

- whether people developed CNV,
- which treatments they received,
- how many injections they needed, and
- how their vision changed over time.

What did they find?

Most people with PXE developed CNV

- **79% of eyes** developed CNV.
- **70% of those eyes** received anti-VEGF injections.

This confirms that CNV is a very common complication of PXE.

Most people needed ongoing treatment

Treatment did **not** stop after the first year.

On average, treated eyes received:

- about **6 injections during the first year**
- about **5 injections during the third year**
- about **6 injections during the fifth year**

In other words, many people continued needing injections year after year.

Vision was generally preserved

Among eyes that received treatment:

- vision decreased by only **1 letter after 1 year**
- **4 letters after 3 years**

- **7 letters after 5 years**

Although vision slowly declined, the average change was relatively modest considering the severity of the disease.

Which medicines were used?

The most commonly used medicines were:

- **Bevacizumab (Avastin)** – 61%
- **Aflibercept (Eylea)** – 43%
- **Ranibizumab (Lucentis)** – 27%

Many people received more than one medication during their care. Because treatment choices changed over time and patients were not randomly assigned to drugs, the study cannot determine whether one medicine works better than another.

What does this mean for people with PXE?

The results are encouraging.

Before anti-VEGF therapy became available, CNV often caused severe, permanent vision loss. This study shows that many people with PXE can now maintain useful vision for years with treatment.

However, there are important limitations:

- Anti-VEGF injections **do not cure PXE.**
- They **do not stop Bruch's membrane from becoming calcified.**
- They **do not prevent new CNV from developing.**
- Many people continue to need injections for years.

The treatment controls one complication of PXE—it does not treat the underlying disease.

What were the study's limitations?

The study looked back at existing medical records rather than following people according to a research protocol.

Because the data came from retina specialists, the study probably included more people with serious eye disease than the overall PXE population. That means the reported **79% rate of CNV may be higher than the true lifetime risk for all people with PXE.** The researchers also could not determine why some people did not receive injections or directly compare the effectiveness of different drugs.

Bottom line

Anti-VEGF injections have transformed the outlook for people with PXE who develop CNV. Most people who receive treatment maintain useful vision for many years, although they usually need repeated injections and still experience a gradual decline in vision over time. The study reinforces the importance of regular retinal monitoring and prompt treatment when CNV develops, while also highlighting the continuing need for therapies that address the underlying cause of PXE rather than just one of its complications.

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