Q

For each statement, identify whether it applies to **gyrate atrophy** *or* **choroideremia** *(or* both*, or* neither*)*

- Is a choroidal dystrophy:

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Why all the prevaricating?

Q/A

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Why all the prevaricating?
When choroideremia and gyrate atrophy were first identified, they were categorized as choroidal dystrophies based on their clinical appearance. And the primary site of pathology in gyrate locates to the RPE and choroid, so it is probably fair to call it a choroidal dystrophy of sorts. However, it is now know that the fundamental pathology in choroideremia is that of a [three words, first two hyphenated]. Because of this, choroideremia was considered to be a form of [two words, and their abb.].

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This was the state of play in the BCSC *Retina* book--that is, until publication of the latest revision (the 2018-19 edition). In this edition, the Academy seems to be phasing out the term *retinitis pigmentosa*. (The book states the term is “no longer preferred.”) Further, the scope of conditions covered by this ‘non-preferred’ umbrella term is shrinking. And one of the no-longer-considered-RP conditions is…choroideremia.

tl;dr I don’t know if choroideremia is considered a choroidal dystrophy. Caveat emptor.

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Regarding phenotype, three classic manifestations of RP are absent in choroideremia. What are they?
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Regarding phenotype, three classic manifestations of RP are absent in choroideremia. What are they?
--**Waxy** (two words)
--**Retinal arteriolar** (one word)
--The presence of (two words)

tl;dr I don't know if choroideremia is considered a choroidal dystrophy. Caveat emptor.

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Regarding phenotype, three classic manifestations of RP are absent in choroideremia. What are they?
--**Waxy disc pallor** (the ONH is normal in choroideremia)
--**Retinal arteriolar attenuation** (the retinal arterioles are normal in choroideremia)
--The presence of **bony spicules** (these are absent in choroideremia)

tl;dr I don't know if choroideremia is considered a choroidal dystrophy. Caveat emptor.

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Why are excess ornithine levels a problem?

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Why are excess ornithine levels a problem?
Because ornithine is toxic to the RPE and choroid

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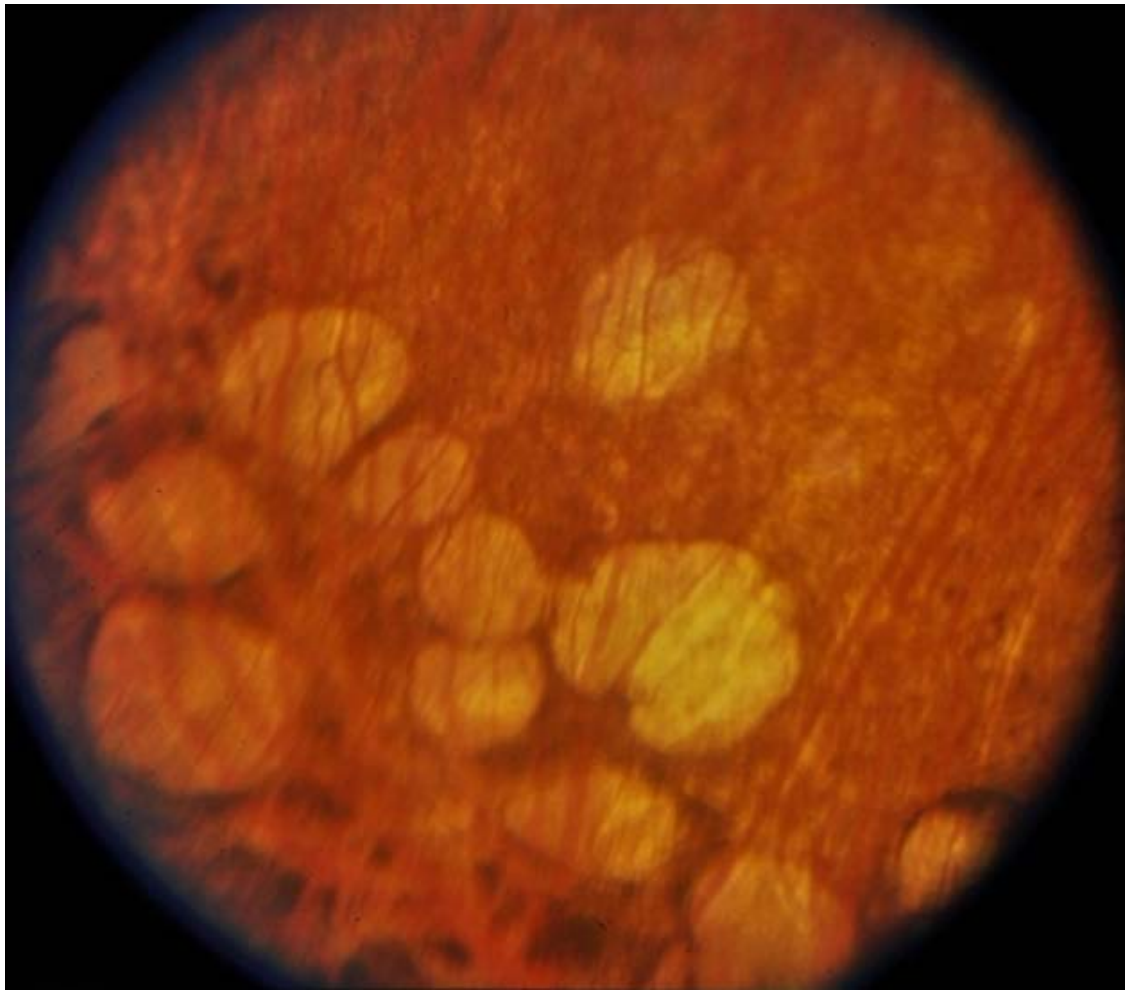

Gyrate atrophy: Pavingstones

For each statement, identify whether it applies to **gyrate atrophy** *or* **choroideremia** *(or* both, *or* neither*)*

Gyrate atrophy: Scalloped areas

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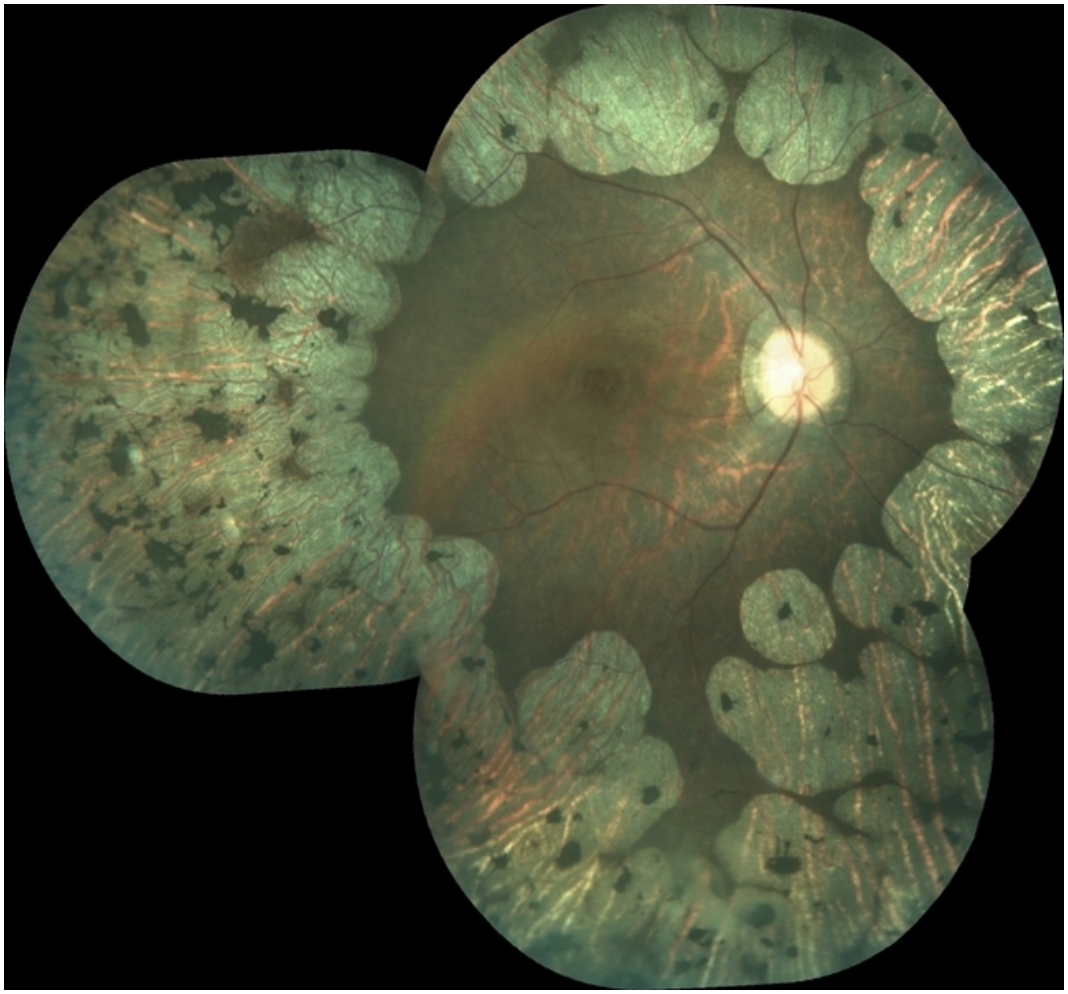

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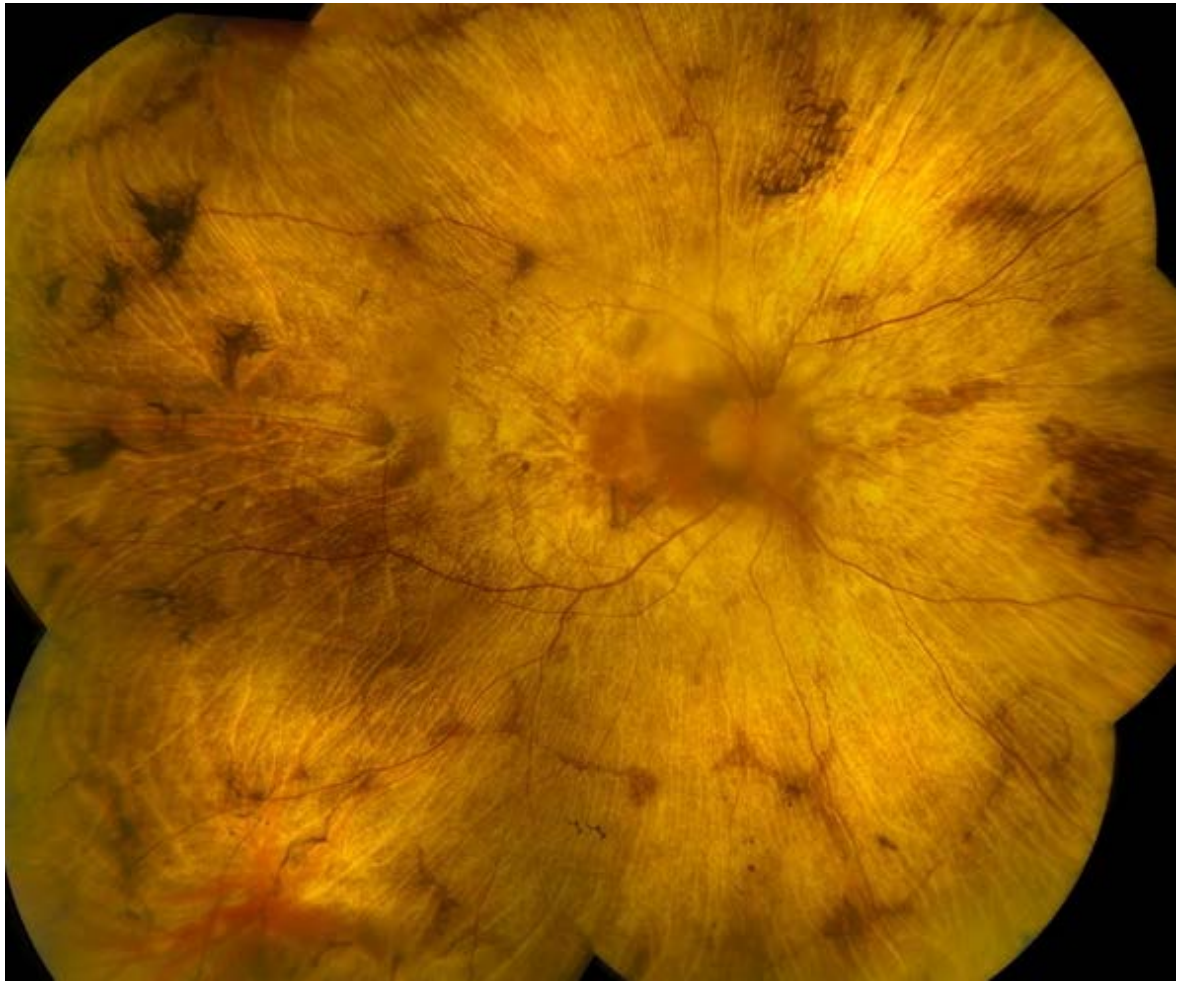

Choroideremia

Q

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- Vitamin B_6 treatment effective in a small percentage of patients:

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If B_6 therapy is tried, check serum [redacted] level to assess response; if level doesn't fall, [redacted] do this

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If B_6 therapy is tried, check serum ornithine level to assess response; if level doesn't fall, discontinue therapy

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- Progressive loss of VF occurs:

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- Treatment: Restrict dietary arginine:

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- Treatment: Restrict dietary arginine: Gyrate