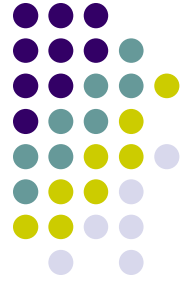


Q

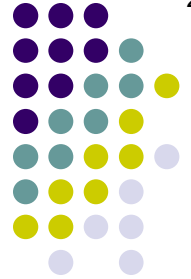
Corneal Ectasias



Briefly, what is a corneal ectasia?

A

Corneal Ectasias



Briefly, what is a corneal ectasia?

To my frustration, I have been unable to locate an 'official' definition in the *BCSC* (or *EyeWiki*, or Krachmer, or Albert & Jakobiec), so the following is based on my review of the aforementioned sources:

A

Corneal Ectasias



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Q

Corneal Ectasias



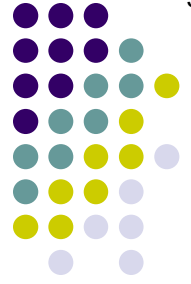
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In comparison, what is a corneal dystrophy?

A

Corneal Ectasias

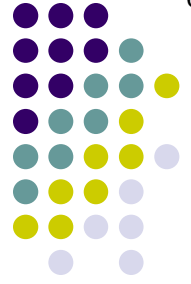


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In comparison, what is a corneal dystrophy?

A dystrophy is an inherited condition characterized by bilateral symmetric changes independent of environmental or systemic processes



Q

Corneal Ectasias

Briefly, what is a corneal ectasia?

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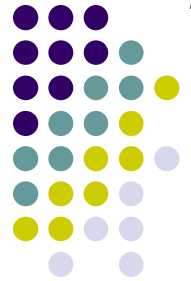
In comparison, what is a corneal dystrophy?

A dystrophy is **an inherited condition characterized by bilateral symmetric changes independent of environmental or systemic processes**

How is that different from an ectasia?

A

Corneal Ectasias



Briefly, what is a corneal ectasia?

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In comparison, what is a corneal dystrophy?

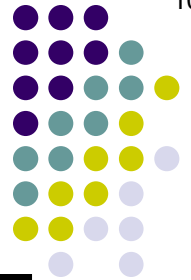
A dystrophy is **an inherited condition characterized by bilateral symmetric changes independent of environmental or systemic processes**

How is that different from an ectasia?

Ectasias are often subject to changes 2ndry to environmental and/or systemic processes

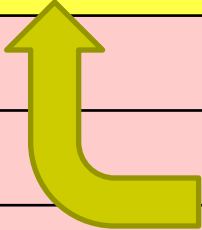
Corneal Ectasias

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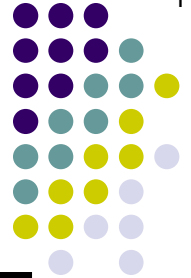
Q

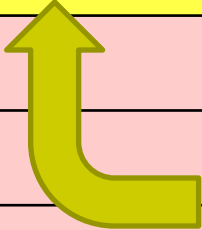
Corneal Ectasias

?	KCN	Keratoglobus	Pellucid Marginal Degeneration
	<p><i>Before we get any further into these three, we would be remiss not to at least mention a fourth (category of) ectasia covered in multiple volumes of the BCSC—remiss because of its importance as a clinical entity (in terms of both its incidence and its impact on vision). What is it?</i></p>		

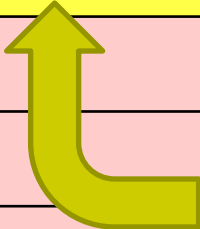
Q/A

Corneal Ectasias



?	KCN	Keratoglobus	Pellucid Marginal Degeneration
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Corneal Ectasias

Post-keratorefractive surgery	KCN	Keratoglobus	Pellucid Marginal Degeneration
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Corneal Ectasias

[illegible]

Corneal Ectasias

What are the risk factors for post-LASIK ectasia?

- KCN or forme fruste KCN
- Thin post-op stromal bed
- Thin pre-op central corneal thickness
- High myopia
- Age <30 years



Corneal Ectasias

Post-keratorefractive surgery	KCN	Keratoglobus	Pellucid Marginal Degeneration
What are the risk factors for post-LASIK ectasia? --KCN or forme fruste KCN			
For more on post-LASIK ectasia, see slide-set RS2			
--High myopia --Age <30 years			

Corneal Ectasias

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Corneal Ectasias

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Corneal Ectasias

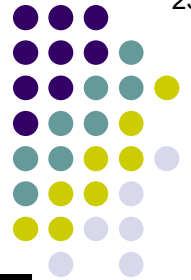
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Corneal Ectasias

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Corneal Ectasias

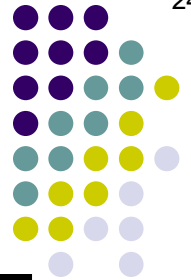
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Corneal Ectasias

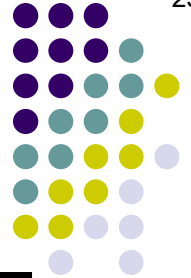
	KCN	Keratoglobus	Pellucid Marginal Degeneration
<i>How common?</i>	Quite	Rare	Uncommon
<i>Age at onset</i>	Teens	Birth	20-40
<i>Thins where?</i>	Inferior paracentral	Global, especially peripheral	Inferior peripheral strip



Q

Corneal Ectasias

	KCN	Keratoglobus	Pellucid Marginal Degeneration
<i>How common?</i>	Quite	Rare	Uncommon
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<i>Thins where?</i>	Inferior paracentral	Global, especially peripheral	Inferior peripheral strip
<i>Protrudes where?</i>			



A

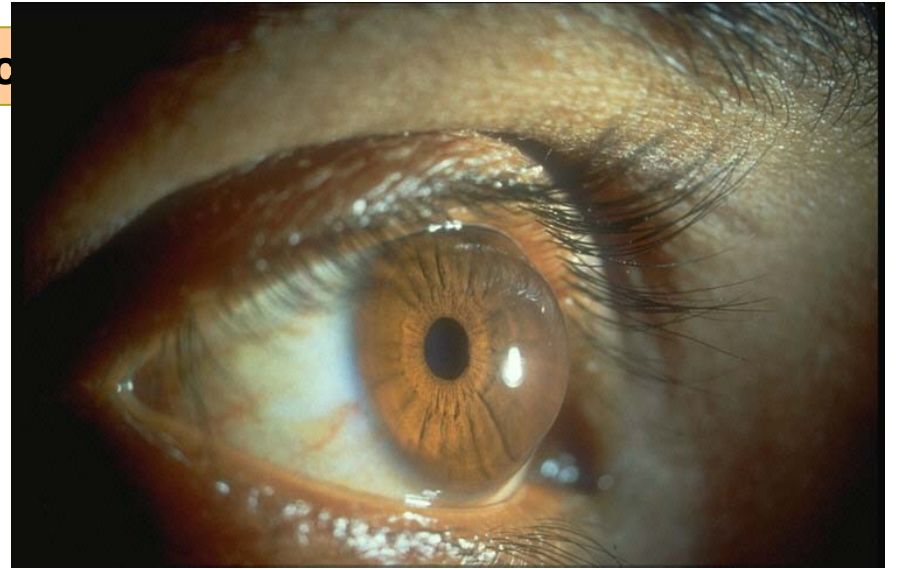
Corneal Ectasias

	KCN	Keratoglobus	Pellucid Marginal Degeneration
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<i>Protrudes where?</i>	At thinning	Global	Above thinning

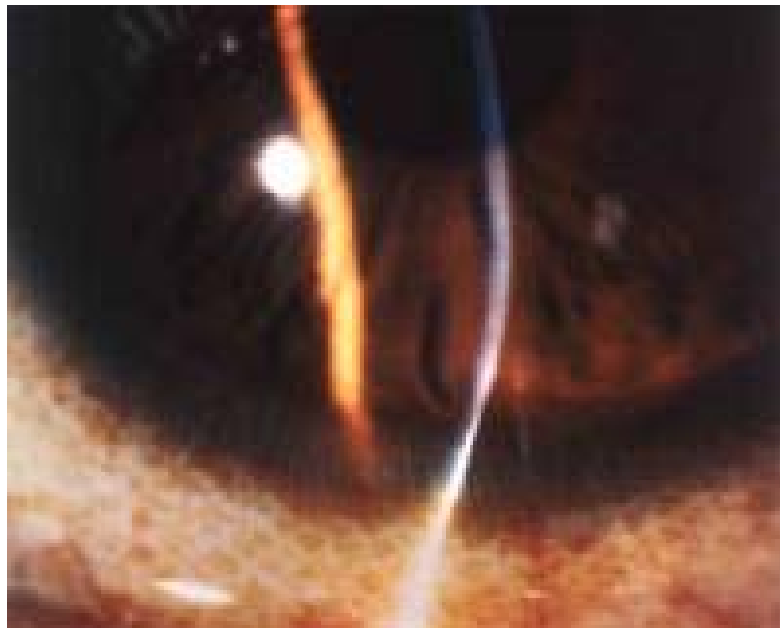
Real Eye



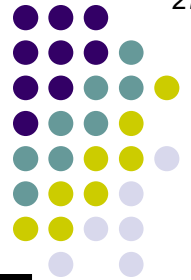
KCN: Protrudes at thinned area



Keratoglobus: Protrudes globally



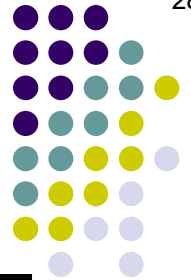
Pellucid: Protrudes *above* thinned area



Corneal Ectasias

	KCN	Keratoglobus	Pellucid Marginal Degeneration
<i>How common?</i>	Quite	Rare	Uncommon
<i>Age at onset</i>	Teens	Birth	20-40
<i>Thins where?</i>	Inferior paracentral	Global, especially peripheral	Inferior peripheral strip
<i>Protrudes where?</i>	At thinning	Global	Above thinning

Make sure you take note of this! It is literally the only ectasia-related factoid that authors of the *Cornea* book felt was worthy of mention in the *Highlights* section of the ectasia/dystrophy chapter!



Q

Corneal Ectasias

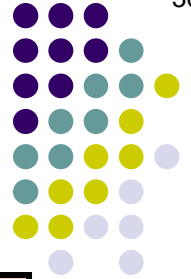
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<i>Thins where?</i>	Inferior paracentral	Global, especially peripheral	Inferior peripheral strip
<i>Protrudes where?</i>	At thinning	Global	Above thinning
<i>Iron line present?</i>			



A

Corneal Ectasias

	KCN	Keratoglobus	Pellucid Marginal Degeneration
<i>How common?</i>	Quite	Rare	Uncommon
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<i>Thins where?</i>	Inferior paracentral	Global, especially peripheral	Inferior peripheral strip
<i>Protrudes where?</i>	At thinning	Global	Above thinning
<i>Iron line present?</i>	Yes	No	Sometimes



Q

Corneal Ectasias

	KCN	Keratoglobus	Pellucid Marginal Degeneration
What is the eponymous name of the iron line in KCN?			Uncommon
			20-40
			Inferior peripheral strip
			Above thinning
Iron line present?	Yes	No	Sometimes

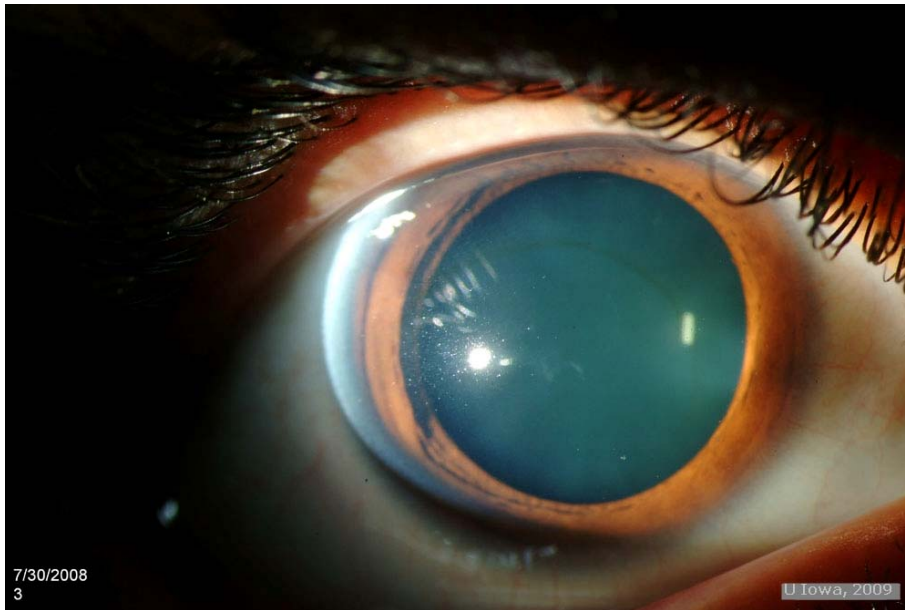
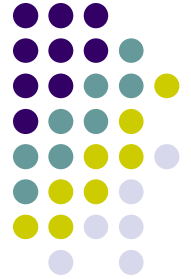


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Corneal Ectasias

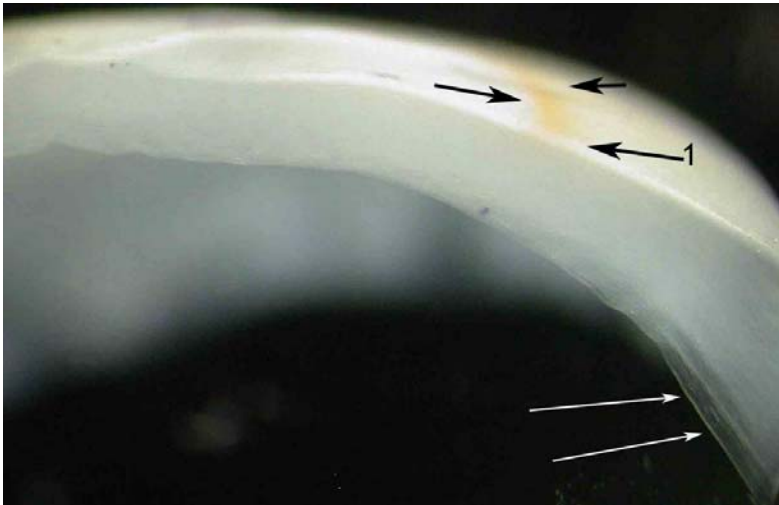
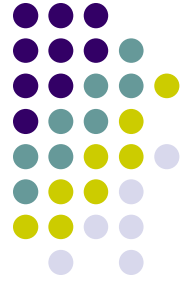
	KCN	Keratoglobus	Pellucid Marginal Degeneration
What is the eponymous name of the iron line in KCN? Fleischer ring			Uncommon
			20-40
			Inferior peripheral strip
			Above thinning
Iron line present?	Yes	No	Sometimes

Corneal Ectasias



KCN: Fleischer ring

Corneal Ectasias



Gross photo. The brown pigment is oxidized iron within the epithelium, constituting the Fleischer ring. (The white arrows show Descemet's membrane.)

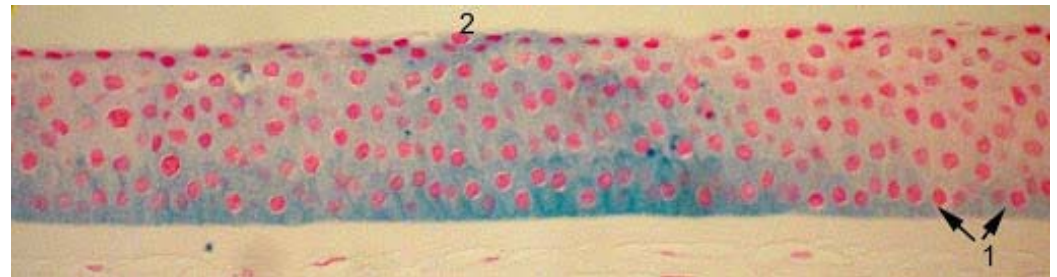
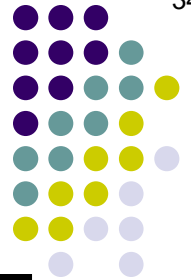


Image shows iron (blue staining at 1) in the basal layer of the epithelium, which transitions to involve the entire epithelium in long standing and severe cases (2).

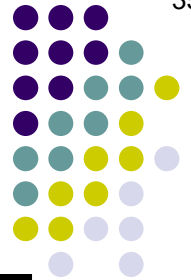
KCN: Fleischer ring



Q

Corneal Ectasias

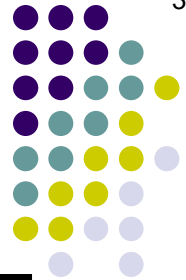
	KCN	Keratoglobus	Pellucid Marginal Degeneration
What is the eponymous name of the iron line in KCN?	Fleischer ring		Uncommon
Where (relative to the cone) is the Fleischer ring typically found?			20-40
			Inferior peripheral strip
			Above thinning
Iron line present?			Sometimes



A

Corneal Ectasias

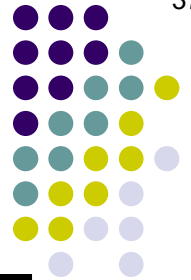
	KCN	Keratoglobus	Pellucid Marginal Degeneration
What is the eponymous name of the iron line in KCN? Fleischer ring			Uncommon
Where (relative to the cone) is the Fleischer ring typically found? Along its inferior extent			20-40
			Inferior peripheral strip
			Above thinning
Iron line present?	Yes	No	Sometimes



Q

Corneal Ectasias

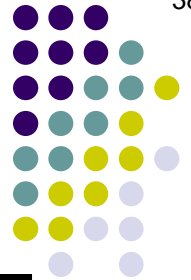
	KCN	Keratoglobus	Pellucid Marginal Degeneration
What is the eponymous name of the iron line in KCN? Fleischer ring			Uncommon
Where (relative to the cone) is the Fleischer ring typically found? Along its inferior extent			20-40
What simple slit-lamp exam maneuver can one do to enhance the visibility of a Fleischer ring?			Inferior peripheral strip
			Above thinning
Iron line present?	Yes	No	Sometimes



A

Corneal Ectasias

	KCN	Keratoglobus	Pellucid Marginal Degeneration
What is the eponymous name of the iron line in KCN? Fleischer ring			Uncommon
Where (relative to the cone) is the Fleischer ring typically found? Along its inferior extent			20-40
What simple slit-lamp exam maneuver can one do to enhance the visibility of a Fleischer ring? Examine the cornea using the cobalt-blue light			Inferior peripheral strip Above thinning
Iron line present?	Yes	No	Sometimes



Q

Corneal Ectasias

	KCN	Keratoglobus	Pellucid Marginal Degeneration
<i>How common?</i>	Quite	Rare	Uncommon
<i>Age at onset</i>	Teens	Birth	20-40
<i>Thins where?</i>	Inferior paracentral	Global, especially peripheral	Inferior peripheral strip
<i>Protrudes where?</i>	At thinning	Global	Above thinning
<i>Iron line present?</i>	Yes	No	Sometimes
<i>Spontaneous rupture?</i>			



A

Corneal Ectasias

	KCN	Keratoglobus	Pellucid Marginal Degeneration
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<i>Protrudes where?</i>	At thinning	Global	Above thinning
<i>Iron line present?</i>	Yes	No	Sometimes
<i>Spontaneous rupture?</i>	Very rare	Very common	Less rare



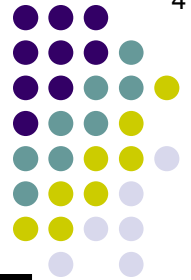
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Corneal Ectasias

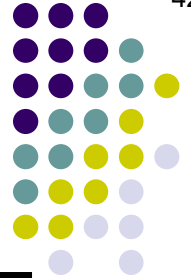
	KCN	Keratoglobus	Pellucid Marginal Degeneration
<i>How common?</i>	Quite	Rare	Uncommon
<i>Age</i>	In the present context, what is acute hydrops?		
<i>Thin</i>			
<i>Protrusion</i>			
<i>Iron line</i>			
<i>Spontaneous rupture?</i>			
	very rare	very common	Less rare

Q/A

Corneal Ectasias



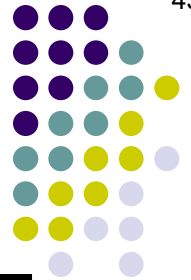
	KCN	Keratoglobus	Pellucid Marginal Degeneration
<i>How common?</i>	Quite	Rare	Uncommon
<i>Age</i>	<p><i>In the present context, what is acute hydrops?</i></p> <p>The sudden development of severe two words 2ndry to a break in eponym</p>		
<i>Thin</i>			
<i>Protrusion</i>			
<i>Iron line</i>			
<i>Spontaneous rupture?</i>			
	very rare	very common	Less rare



A

Corneal Ectasias

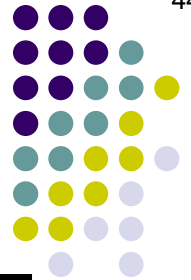
	KCN	Keratoglobus	Pellucid Marginal Degeneration
<i>How common?</i>	Quite	Rare	Uncommon
<i>Age</i>	<p><i>In the present context, what is acute hydrops?</i> The sudden development of severe corneal edema 2ndry to a break in Descemet's</p>		
<i>Thin</i>			
<i>Protrusion</i>			
<i>Iron line</i>			
<i>Spontaneous rupture?</i>			
	very rare	very common	Less rare



Q

Corneal Ectasias

	KCN	Keratoglobus	Pellucid Marginal Degeneration
<i>How common?</i>	Quite	Rare	Uncommon
<i>Age</i>	<p><i>In the present context, what is acute hydrops?</i> The sudden development of severe corneal edema 2ndry to a break in Descemet's</p> <p><i>With which ectasia is it most closely associated?</i></p>		
<i>Thin</i>			
<i>Protrusion</i>			
<i>Iron line</i>			
<i>Spontaneous rupture?</i>			
	very rare	very common	Less rare

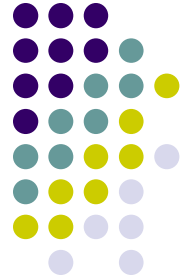


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Corneal Ectasias

	KCN	Keratoglobus	Pellucid Marginal Degeneration
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<i>Thin</i>			
<i>Protrusion</i>			
<i>Iron line</i>			
<i>Spontaneous rupture?</i>			
	very rare	very common	Less rare

Corneal Ectasias



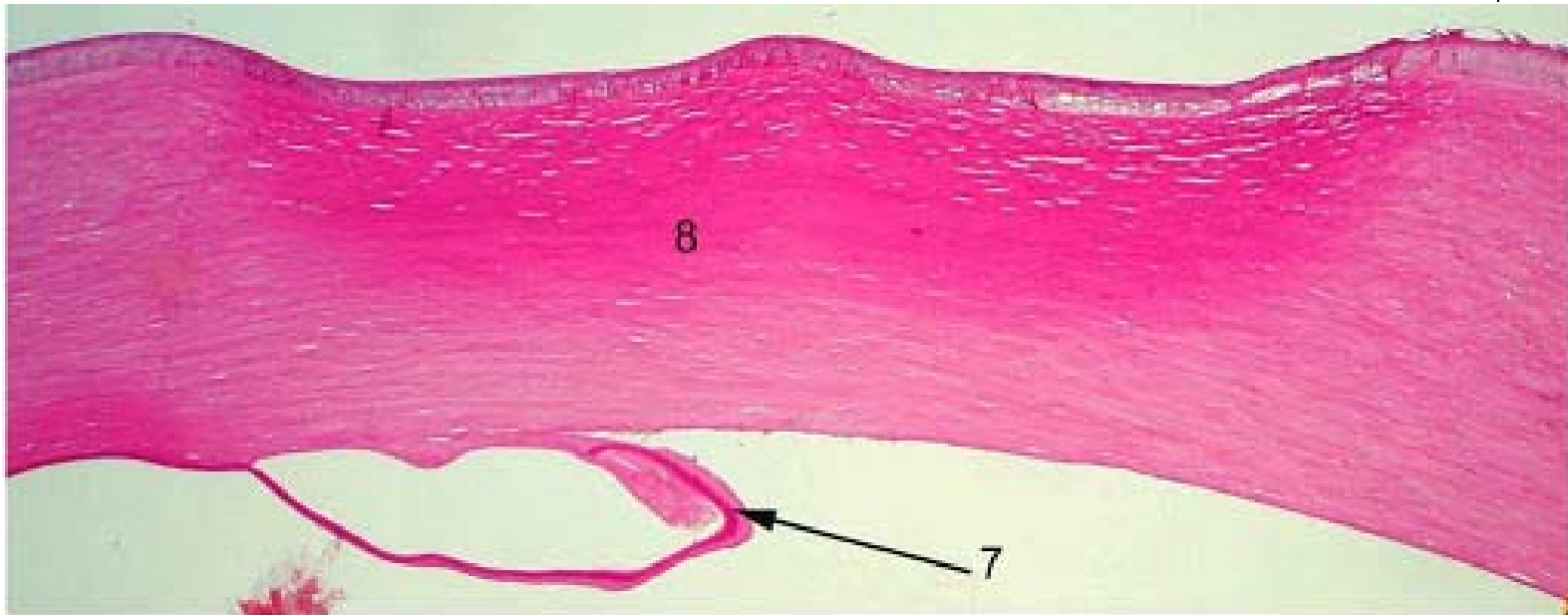
3 days after break



Scarring, same eye, 2 months+ later

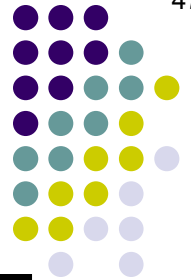
KCN: Acute hydrops

Corneal Ectasias



Corneal hydrops. Descemet's membrane is disrupted and folded back to rejoin the cornea in the wrong orientation (7) . There is marked stromal scarring (8).

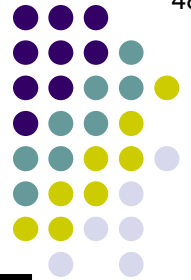
KCN: Acute hydrops



Q

Corneal Ectasias

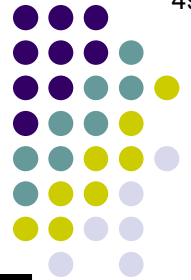
	KCN	Keratoglobus	Pellucid Marginal Degeneration
<i>How common?</i>	Quite	Rare	Uncommon
<i>Age</i>	<p><i>In the present context, what is acute hydrops?</i> The sudden development of severe corneal edema 2ndry to a break in Descemet's</p> <p><i>With which ectasia is it most closely associated?</i> KCN</p> <p><i>Can it occur in keratoglobus and/or PMD?</i></p>		
<i>Thin</i>			
<i>Protrusion</i>			
<i>Iron line</i>			
<i>Spontaneous rupture?</i>			
	very rare	very common	Less rare



A

Corneal Ectasias

	KCN	Keratoglobus	Pellucid Marginal Degeneration
<i>How common?</i>	Quite	Rare	Uncommon
<i>Age</i>	<p><i>In the present context, what is acute hydrops?</i> The sudden development of severe corneal edema 2ndry to a break in Descemet's</p> <p><i>With which ectasia is it most closely associated?</i> KCN</p> <p><i>Can it occur in keratoglobus and/or PMD?</i> Yes, but it is much less likely</p>		
<i>Thin</i>			
<i>Protrusion</i>			
<i>Iron line</i>			
<i>Spontaneous rupture?</i>			
	very rare	very common	Less rare



Q

Corneal Ectasias

	KCN	Keratoglobus	Pellucid Marginal Degeneration
<i>How common?</i>	Quite	Rare	Uncommon
<i>Age at onset</i>	Teens	Birth	20-40
<i>Thins where?</i>	Inferior paracentral	Global, especially peripheral	Inferior peripheral strip
<i>Protrudes where?</i>	At thinning	Global	Above thinning
<i>Iron line present?</i>	Yes	No	Sometimes
<i>Spontaneous rupture?</i>	Very rare	Very common	Less rare
<i>Steepens where?</i>			



A

Corneal Ectasias

	KCN	Keratoglobus	Pellucid Marginal Degeneration
<i>How common?</i>	Quite	Rare	Uncommon
<i>Age at onset</i>	Teens	Birth	20-40
<i>Thins where?</i>	Inferior paracentral	Global, especially peripheral	Inferior peripheral strip
<i>Protrudes where?</i>	At thinning	Global	Above thinning
<i>Iron line present?</i>	Yes	No	Sometimes
<i>Spontaneous rupture?</i>	Very rare	Very common	Less rare
<i>Steepens where?</i>	Below thinning	Globally	Inferiorly



Q

Corneal Ectasias

	KCN	Keratoglobus	Pellucid Marginal Degeneration
<i>How common?</i>	Quite	Rare	Uncommon
<i>Age at onset</i>	Teens	Birth	20-40
<i>Thins where?</i>	Inferior paracentral	Global, especially peripheral	Inferior peripheral strip
<i>Protrudes where?</i>	At thinning	Global	Above thinning
<i>Iron line present?</i>	Yes	No	Sometimes
<i>Spontaneous rupture?</i>	Very rare	Very common	Less rare
<i>Steepens where?</i>	Below thinning	Globally	Inferiorly
<i>Munson sign?</i>			



A

Corneal Ectasias

	KCN	Keratoglobus	Pellucid Marginal Degeneration
<i>How common?</i>	Quite	Rare	Uncommon
<i>Age at onset</i>	Teens	Birth	20-40
<i>Thins where?</i>	Inferior paracentral	Global, especially peripheral	Inferior peripheral strip
<i>Protrudes where?</i>	At thinning	Global	Above thinning
<i>Iron line present?</i>	Yes	No	Sometimes
<i>Spontaneous rupture?</i>	Very rare	Very common	Less rare
<i>Steepens where?</i>	Below thinning	Globally	Inferiorly
<i>Munson sign?</i>	Yes	?Yes?	Yes



Q

Corneal Ectasias

	KCN	Keratoglobus	Pellucid Marginal Degeneration
<i>How common?</i>	Quite	Rare	Uncommon
<i>Age at onset</i>	Teens	Birth	20-40
<i>Thins where?</i>	Inferior paracentral	Global, especially peripheral	Inferior peripheral strip
<i>Protrudes where?</i>	At thinning	Global	Above thinning
<i>Iron line present?</i>	Yes	No	Sometimes
<i>Spontaneous</i>	What is Munson sign?		
<i>Steeper</i>			
Munson sign?	Yes	?Yes?	Yes



A

Corneal Ectasias

	KCN	Keratoglobus	Pellucid Marginal Degeneration
<i>How common?</i>	Quite	Rare	Uncommon
<i>Age at onset</i>	Teens	Birth	20-40
<i>Thins where?</i>	Inferior paracentral	Global, especially peripheral	Inferior peripheral strip
<i>Protrudes where?</i>	At thinning	Global	Above thinning
<i>Iron line present?</i>	Yes	No	Sometimes
<i>Spontaneous</i>	What is Munson sign? Displacement of the central lower lid by the protruding cornea when the eye is in downgaze		
<i>Steeper</i>			
Munson sign?	Yes	?Yes?	Yes

Corneal Ectasias



Munson's sign in KCN



Q

Corneal Ectasias

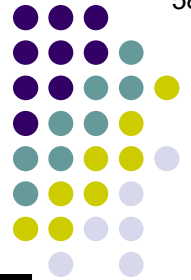
	KCN	Keratoglobus	Pellucid Marginal Degeneration
<i>How common?</i>	<i>Of the three, which has the strongest hereditary component?</i>		
<i>Age at onset</i>			
<i>Thins where?</i>			
<i>Protrudes where?</i>	At thinning	Global	Above thinning
<i>Iron line present?</i>	Yes	No	Sometimes
<i>Spontaneous rupture?</i>	Very rare	Very common	Less rare
<i>Steepens where?</i>	Below thinning	Globally	Inferiorly
<i>Munson sign?</i>	Yes	?Yes?	Yes



A

Corneal Ectasias

	KCN	Keratoglobus	Pellucid Marginal Degeneration
<i>How common?</i>	Of the three, which has the strongest hereditary component? KCN		
<i>Age at onset</i>			
<i>Thins where?</i>			
<i>Protrudes where?</i>	At thinning	Global	Above thinning
<i>Iron line present?</i>	Yes	No	Sometimes
<i>Spontaneous rupture?</i>	Very rare	Very common	Less rare
<i>Steepens where?</i>	Below thinning	Globally	Inferiorly
<i>Munson sign?</i>	Yes	?Yes?	Yes



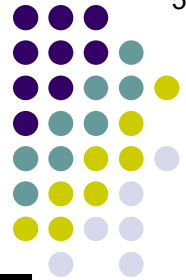
Q

Corneal Ectasias

	KCN	Keratoglobus	Pellucid Marginal Degeneration
<i>How common?</i>	<p>Of the three, which has the strongest hereditary component? KCN</p> <p>How strong is the hereditary component of KCN?</p>		
<i>Age at onset</i>			
<i>Thins where?</i>			
<i>Protrudes where?</i>	At thinning	Global	Above thinning
<i>Iron line present?</i>	Yes	No	Sometimes
<i>Spontaneous rupture?</i>	Very rare	Very common	Less rare
<i>Steepens where?</i>	Below thinning	Globally	Inferiorly
<i>Munson sign?</i>	Yes	?Yes?	Yes

Q/A

Corneal Ectasias



	KCN	Keratoglobus	Pellucid Marginal Degeneration
<i>How common?</i>	<p>Of the three, which has the strongest hereditary component? KCN</p> <p>How strong is the hereditary component of KCN? Not very--family hx is positive in only about % of cases (but that's still higher than in either keratoglobus or PMD)</p>		
<i>Age at onset</i>			
<i>Thins where?</i>			
<i>Protrudes where?</i>	At thinning	Global	Above thinning
<i>Iron line present?</i>	Yes	No	Sometimes
<i>Spontaneous rupture?</i>	Very rare	Very common	Less rare
<i>Steepens where?</i>	Below thinning	Globally	Inferiorly
<i>Munson sign?</i>	Yes	?Yes?	Yes



A

Corneal Ectasias

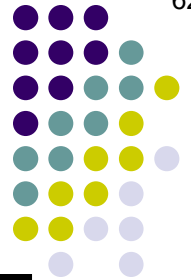
	KCN	Keratoglobus	Pellucid Marginal Degeneration
<i>How common?</i>	<p>Of the three, which has the strongest hereditary component? KCN</p> <p>How strong is the hereditary component of KCN? Not very--family hx is positive in only about 7% of cases (but that's still higher than in either keratoglobus or PMD)</p>		
<i>Age at onset</i>			
<i>Thins where?</i>			
<i>Protrudes where?</i>	At thinning	Global	Above thinning
<i>Iron line present?</i>	Yes	No	Sometimes
<i>Spontaneous rupture?</i>	Very rare	Very common	Less rare
<i>Steepens where?</i>	Below thinning	Globally	Inferiorly
<i>Munson sign?</i>	Yes	?Yes?	Yes



Q

Corneal Ectasias

	KCN	Keratoglobus	Pellucid Marginal Degeneration
How common		Keratoglobus has two strong syndromic associations--what are they? -- --	
Age at onset			
Thins where			
Protrudes where			
Iron line present			
Spontaneous rupture			
Steepens where?	Below thinning	Globally	Inferiorly
Munson sign?	Yes	?Yes?	Yes



A

Corneal Ectasias

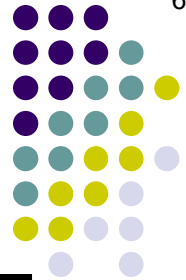
	KCN	Keratoglobus	Pellucid Marginal Degeneration
How common		<p><i>Keratoglobus has two strong syndromic associations--what are they?</i></p> <p>--Osteogenesis imperfecta (OI)</p> <p>--Ehlers-Danlos syndrome (EDS)</p>	
Age at onset			
Thins where			
Protrudes where			
Iron line present			
Spontaneous rupture			
Steepens where?	Below thinning	Globally	Inferiorly
Munson sign?	Yes	?Yes?	Yes



Q

Corneal Ectasias

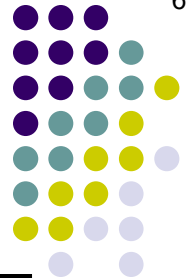
	KCN	Keratoglobus	Pellucid Marginal Degeneration
How common		<p><i>Keratoglobus has two strong syndromic associations--what are they?</i></p> <p>--Osteogenesis imperfecta (OI)</p> <p>--Ehlers-Danlos syndrome (EDS)</p> <p><i>In two, hyphenated words, what sort of disorder are OI and EDS?</i></p> <p>They are disorders</p>	
Age at onset			
Thins where			
Protrudes where			
Iron line present			
Spontaneous rupture			
Steepens where?	Below thinning	Globally	Interiorly
Munson sign?	Yes	?Yes?	Yes



A

Corneal Ectasias

	KCN	Keratoglobus	Pellucid Marginal Degeneration
How common?		<p><i>Keratoglobus has two strong syndromic associations--what are they?</i> --Osteogenesis imperfecta (OI) --Ehlers-Danlos syndrome (EDS)</p> <p><i>In two, hyphenated words, what sort of disorder are OI and EDS?</i> They are connective-tissue disorders</p>	
Age at onset			
Thins where?			
Protrudes where?			
Iron line present?			
Spontaneous rupture?			
Steepens where?	Below thinning	Globally	Inferiorly
Munson sign?	Yes	?Yes?	Yes



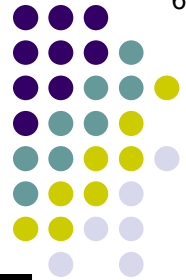
Q

Corneal Ectasias

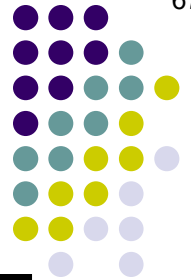
	KCN	Keratoglobus	Pellucid Marginal Degeneration
How common?		<p><i>Keratoglobus has two strong syndromic associations--what are they?</i> --Osteogenesis imperfecta (OI) --Ehlers-Danlos syndrome (EDS)</p> <p><i>In two, hyphenated words, what sort of disorder are OI and EDS?</i> They are connective-tissue disorders</p> <p><i>Briefly, what are the main nonocular findings in each?</i> OI: EDS:</p>	
Age at onset			
Thins where?			
Protrudes where?			
Iron line present?			
Spontaneous rupture?			
Steepens where?	Below thinning	Globally	Inferiorly
Munson sign?	Yes	?Yes?	Yes

Q/A

Corneal Ectasias



	KCN	Keratoglobus	Pellucid Marginal Degeneration
How common?		<p><i>Keratoglobus has two strong syndromic associations--what are they?</i> --Osteogenesis imperfecta (OI) --Ehlers-Danlos syndrome (EDS)</p> <p><i>In two, hyphenated words, what sort of disorder are OI and EDS?</i> They are connective-tissue disorders</p> <p><i>Briefly, what are the main nonocular findings in each?</i> OI: Fragile box; deafness EDS:</p>	
Age at onset			
Thins where?			
Protrudes where?			
Iron line present?			
Spontaneous rupture?			
Steepens where?	Below thinning	Globally	Inferiorly
Munson sign?	Yes	?Yes?	Yes



A

Corneal Ectasias

	KCN	Keratoglobus	Pellucid Marginal Degeneration
How common?		<p><i>Keratoglobus has two strong syndromic associations--what are they?</i> --Osteogenesis imperfecta (OI) --Ehlers-Danlos syndrome (EDS)</p> <p><i>In two, hyphenated words, what sort of disorder are OI and EDS?</i> They are connective-tissue disorders</p> <p><i>Briefly, what are the main nonocular findings in each?</i> OI: Fragile bones ; deafness EDS:</p>	
Age at onset			
Thins where?			
Protrudes where?			
Iron line present?			
Spontaneous rupture?			
Steepens where?	Below thinning	Globally	Inferiorly
Munson sign?	Yes	?Yes?	Yes

Q/A

Corneal Ectasias



	KCN	Keratoglobus	Pellucid Marginal Degeneration
How common?		<p><i>Keratoglobus has two strong syndromic associations--what are they?</i> --Osteogenesis imperfecta (OI) --Ehlers-Danlos syndrome (EDS)</p> <p><i>In two, hyphenated words, what sort of disorder are OI and EDS?</i> They are connective-tissue disorders</p> <p><i>Briefly, what are the main nonocular findings in each?</i> OI: Fragile bones ; deafness EDS: Fragile ; hyperextensible joints</p>	
Age at onset			
Thins where?			
Protrudes where?			
Iron line present?			
Spontaneous rupture?			
Steepens where?	Below thinning	Globally	Inferiorly
Munson sign?	Yes	?Yes?	Yes



A

Corneal Ectasias

	KCN	Keratoglobus	Pellucid Marginal Degeneration
How common		<p><i>Keratoglobus has two strong syndromic associations--what are they?</i> --Osteogenesis imperfecta (OI) --Ehlers-Danlos syndrome (EDS)</p> <p><i>In two, hyphenated words, what sort of disorder are OI and EDS?</i> They are connective-tissue disorders</p> <p><i>Briefly, what are the main nonocular findings in each?</i> OI: Fragile bones ; deafness EDS: Fragile skin ; hyperextensible joints</p>	
Age at onset			
Thins where			
Protrudes where			
Iron line present			
Spontaneous rupture			
Steepens where?	Below thinning	Globally	Interiorly
Munson sign?	Yes	?Yes?	Yes

Corneal Ectasias



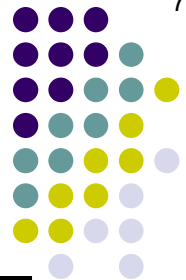
Ehlers-Danlos syndrome



Q

Corneal Ectasias

	KCN	Keratoglobus	Pellucid Marginal Degeneration
How common	<p><i>Keratoglobus has two strong syndromic associations--what are they?</i></p> <p>--Osteogenesis imperfecta (OI)</p> <p>--Ehlers-Danlos syndrome (EDS)</p> <p><i>In two, hyphenated words, what sort of disorder are OI and EDS?</i></p> <p>They are connective-tissue disorders</p> <p><i>Briefly, what are the main nonocular findings in each?</i></p> <p>OI: Fragile bones ; deafness; easy...</p> <p>EDS: Fragile skin ; hyperextensible joints; easy...</p> <p><i>They share another finding related to vascular manifestations of abnormal connective tissue--what is it?</i></p>		
Age at onset			
Thins where			
Protrudes where			
Iron line present			
Steepens where?	Below thinning	Globally	Interiorly
Munson sign?	Yes	?Yes?	Yes



A

Corneal Ectasias

	KCN	Keratoglobus	Pellucid Marginal Degeneration
How common?	<p>Keratoglobus has two strong syndromic associations--what are they?</p> <p>--Osteogenesis imperfecta (OI)</p> <p>--Ehlers-Danlos syndrome (EDS)</p> <p>In two, hyphenated words, what sort of disorder are OI and EDS?</p> <p>They are connective-tissue disorders</p> <p>Briefly, what are the main nonocular findings in each?</p> <p>OI: Fragile bones ; deafness; easy...bruising/bleeding</p> <p>EDS: Fragile skin ; hyperextensible joints; easy...bruising/bleeding</p> <p>They share another finding related to vascular manifestations of abnormal connective tissue--what is it?</p> <p>Easy bruising/bleeding</p>		
Age at onset			
Thins where?			
Protrudes where?			
Iron line present?			
Steepens where?	Below thinning	Globally	Interiorly
Munson sign?	Yes	?Yes?	Yes



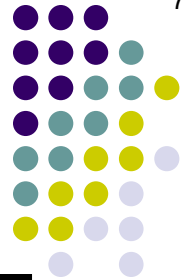
Q

Corneal Ectasias

	KCN	Keratoglobus	Pellucid Marginal Degeneration
How common?		<p><i>Keratoglobus has two strong syndromic associations--what are they?</i> --Osteogenesis imperfecta (OI) --Ehlers-Danlos syndrome (EDS)</p> <p><i>In two, hyphenated words, what sort of disorder are OI and EDS?</i> They are connective-tissue disorders</p> <p><i>Briefly, what are the main nonocular findings in each?</i> OI: Fragile bones ; deafness; easy...bruising/bleeding EDS: Fragile skin ; hyperextensible joints; easy...bruising/bleeding</p> <p><i>What colorful eye finding is associated with both OI and EDS?</i></p>	
Age at onset			
Thins where?			
Protrudes where?			
Iron line present?			
Spontaneous rupture?		<p><i>Below thinning</i></p> <p><i>Globally</i></p> <p><i>Interiorly</i></p>	
Steepens where?			
Munson sign?	Yes	?Yes?	Yes

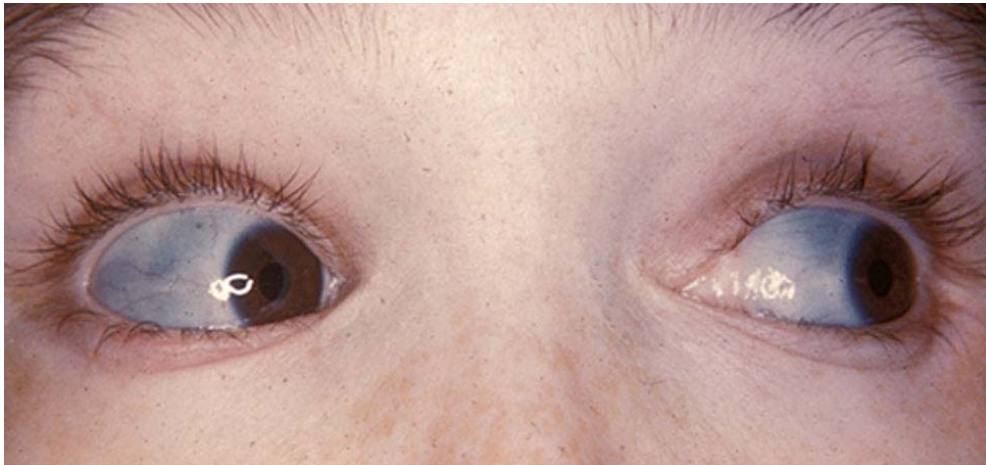
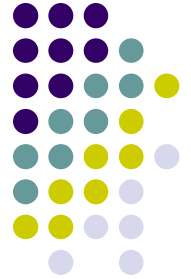
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Corneal Ectasias

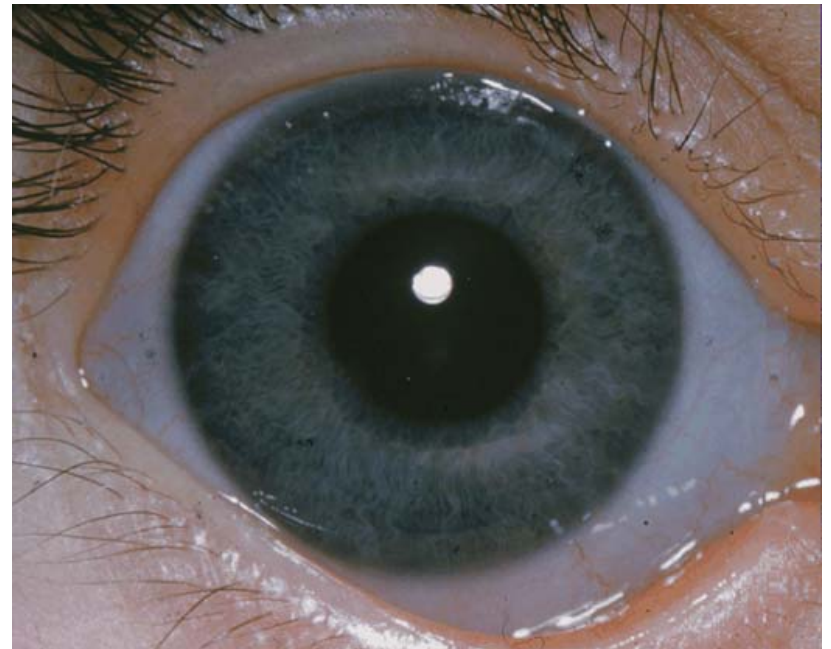


	KCN	Keratoglobus	Pellucid Marginal Degeneration
How common		<p><i>Keratoglobus has two strong syndromic associations--what are they?</i> --Osteogenesis imperfecta (OI) --Ehlers-Danlos syndrome (EDS)</p> <p><i>In two, hyphenated words, what sort of disorder are OI and EDS?</i> They are connective-tissue disorders</p> <p><i>Briefly, what are the main nonocular findings in each?</i> OI: Fragile bones ; deafness; easy...bruising/bleeding EDS: Fragile skin ; hyperextensible joints; easy...bruising/bleeding</p> <p><i>What colorful eye finding is associated with both OI and EDS?</i> Blue sclera</p>	
Age at onset			
Thins where			
Protrudes where			
Iron line present			
Spontaneous rupture		<p><i>Below thinning</i></p> <p><i>Globally</i></p> <p><i>Interiorly</i></p>	
Steepens where?			
Munson sign?	Yes	?Yes?	Yes

Corneal Ectasias



Blue sclera in OI



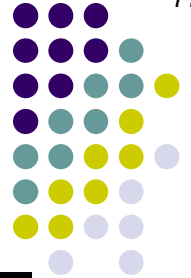
Blue sclera in Ehlers-Danlos



Q

Corneal Ectasias

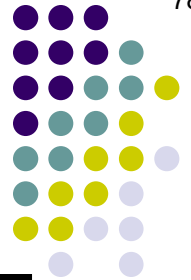
	KCN	Keratoglobus	Pellucid Marginal Degeneration
How common		Keratoglobus has two strong syndromic associations--what are they?	
Age at onset		<ul style="list-style-type: none"> -Osteogenesis imperfecta (OI) -Ehlers-Danlos syndrome (EDS) 	
Thins where		What is the classic posterior-pole association for Ehlers-Danlos?	
Protrudes where			
Iron line present			
Spontaneous rupture			
Steepens where			
Munson sign?	Yes	?Yes?	Yes



A

Corneal Ectasias

	KCN	Keratoglobus	Pellucid Marginal Degeneration
How common		Keratoglobus has two strong syndromic associations--what are they?	
Age at onset		<ul style="list-style-type: none"> -Osteogenesis imperfecta (OI) -Ehlers-Danlos syndrome (EDS) 	
Thins where		What is the classic posterior-pole association for Ehlers-Danlos? Angioid streaks	
Protrudes where			
Iron line present			
Spontaneous rupture			
Steepens where			
Munson sign?	Yes	?Yes?	Yes



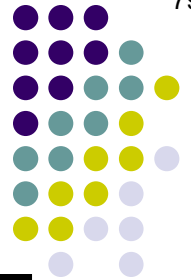
Q

Corneal Ectasias

	KCN	Keratoglobus	Pellucid Marginal Degeneration
How common		Keratoglobus has two strong syndromic associations--what are they?	
Age at onset		<ul style="list-style-type: none"> - Osteogenesis imperfecta (OI) - Ehlers-Danlos syndrome (EDS) 	
Thins where		What is the classic posterior-pole association for Ehlers-Danlos?	
Protrudes where		Angioid streaks	
Iron line present		What is the DFE appearance of angioid streaks?	
Spontaneous rupture			
Steepens where			
Munson sign?	Yes	?Yes?	Yes

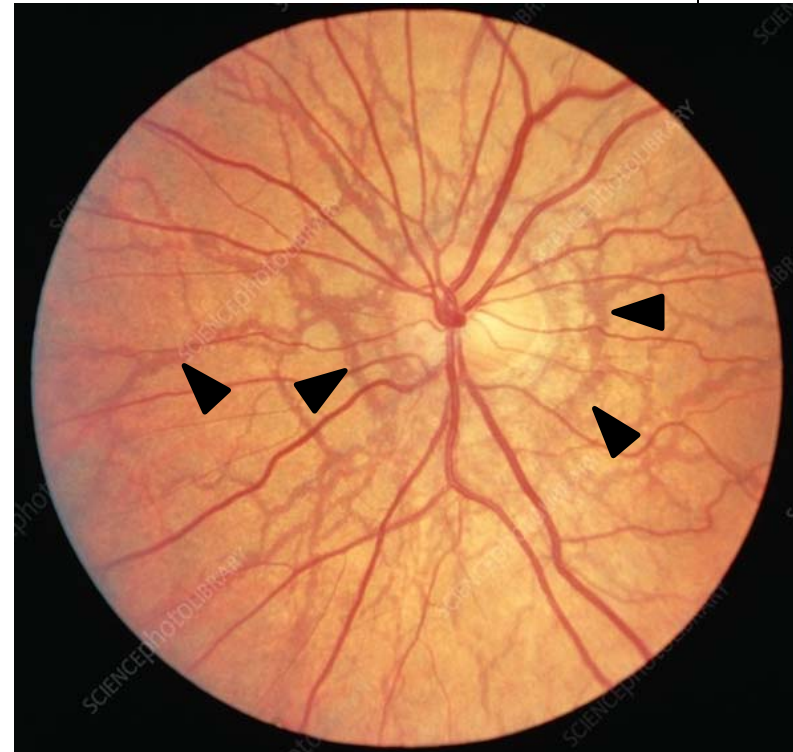
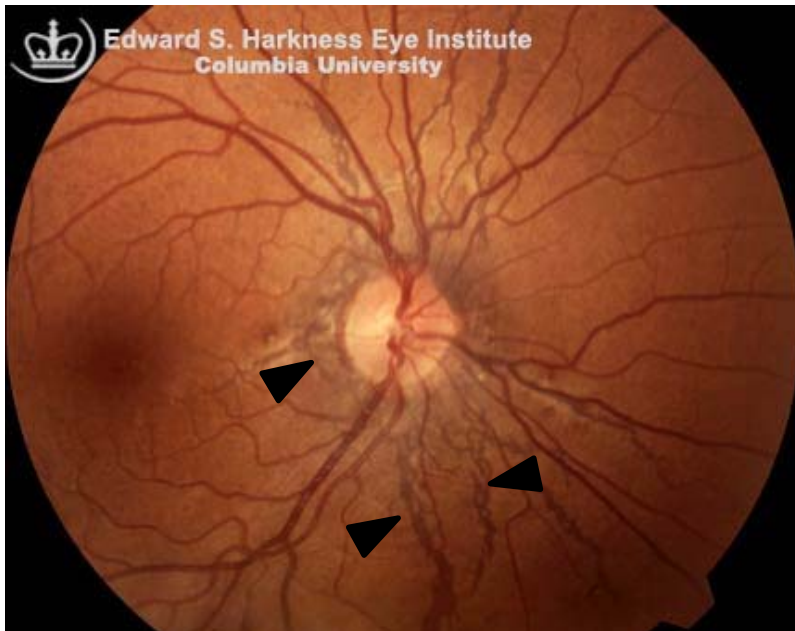
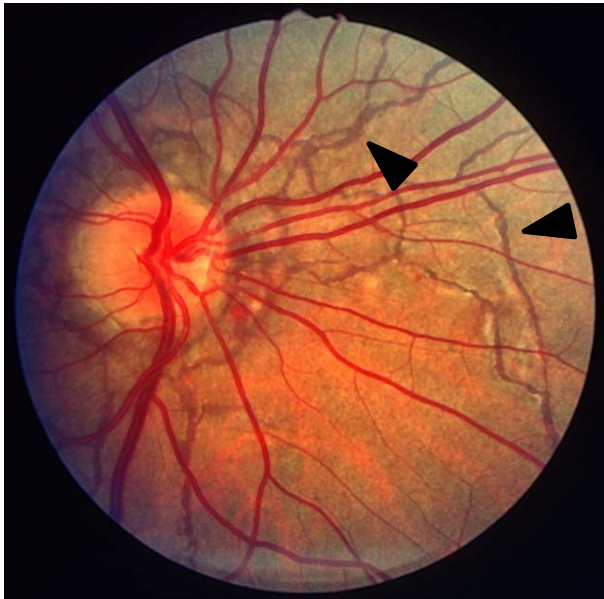
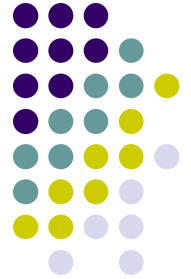
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Corneal Ectasias

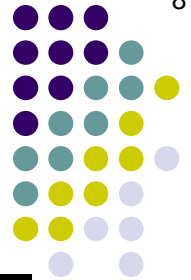


	KCN	Keratoglobus	Pellucid Marginal Degeneration
How common		Keratoglobus has two strong syndromic associations--what are they?	
Age at onset		<ul style="list-style-type: none"> - Osteogenesis imperfecta (OI) - Ehlers-Danlos syndrome (EDS) 	
Thins where		<p>What is the classic posterior-pole association for Ehlers-Danlos?</p> <p>Angioid streaks</p>	
Protrudes where		<p>What is the DFE appearance of angioid streaks?</p> <p>Reddish-brown lines radiating out from the peripapillary region</p>	
Iron line present			
Spontaneous rupture			
Steepens where			
Munson sign?	Yes	?Yes?	Yes

Corneal Ectasias



Angioid streaks (arrowheads).
Note that only a few of the many
present have been marked.



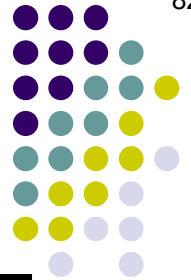
Q

Corneal Ectasias

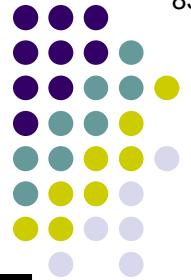
	KCN	Keratoglobus	Pellucid Marginal Degeneration
How common		Keratoglobus has two strong syndromic associations--what are they?	
Age at onset		<ul style="list-style-type: none"> -Osteogenesis imperfecta (OI) -Ehlers-Danlos syndrome (EDS) 	
Thins where		What is the classic posterior-pole association for Ehlers-Danlos? Angioid streaks	
Protrudes where		What is the DFE appearance of angioid streaks? Reddish-brown lines radiating out from the peripapillary region	
Iron line present		Why are they called 'angioid' streaks?	
Spontaneous rupture			
Steepens where			
Munson sign?	Yes	?Yes?	Yes

A

Corneal Ectasias



	KCN	Keratoglobus	Pellucid Marginal Degeneration
How common		Keratoglobus has two strong syndromic associations--what are they?	
Age at onset		<ul style="list-style-type: none"> - Osteogenesis imperfecta (OI) - Ehlers-Danlos syndrome (EDS) 	
Thins where		<p>What is the classic posterior-pole association for Ehlers-Danlos?</p> <p>Angioid streaks</p>	
Protrudes where		<p>What is the DFE appearance of angioid streaks?</p> <p>Reddish-brown lines radiating out from the peripapillary region</p>	
Iron line present		<p>Why are they called 'angioid' streaks?</p> <p>Because the streaks resemble blood vessels</p>	
Spontaneous rupture			
Steepens where			
Munson sign?	Yes	?Yes?	Yes



Q

Corneal Ectasias

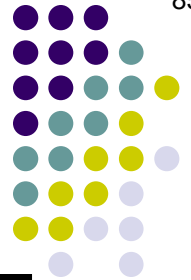
	KCN	Keratoglobus	Pellucid Marginal Degeneration
How common		Keratoglobus has two strong syndromic associations--what are they?	
Age at onset		<ul style="list-style-type: none"> - Osteogenesis imperfecta (OI) - Ehlers-Danlos syndrome (EDS) 	
Thins where		<p>What is the classic posterior-pole association for Ehlers-Danlos?</p> <p>Angioid streaks</p>	
Protrudes where		<p>What is the DFE appearance of angioid streaks?</p> <p>Reddish-brown lines radiating out from the peripapillary region</p>	
Iron line present		<p>Why are they called 'angioid' streaks?</p> <p>Because the streaks resemble blood vessels</p>	
Spontaneous rupture		<p>Which tissue is the abnormal in angioid streaks?</p>	
Steepens where			
Munson sign?	Yes	?Yes?	Yes

A

Corneal Ectasias



	KCN	Keratoglobus	Pellucid Marginal Degeneration
How common		Keratoglobus has two strong syndromic associations--what are they?	
Age at onset		<ul style="list-style-type: none"> -Osteogenesis imperfecta (OI) -Ehlers-Danlos syndrome (EDS) 	
Thins where		What is the classic posterior-pole association for Ehlers-Danlos?	
Protrudes where		Angioid streaks	
Iron line present		What is the DFE appearance of angioid streaks?	
Spontaneous rupture		Reddish-brown lines radiating out from the peripapillary region	
Steepens where		Why are they called 'angioid' streaks?	
		Because the streaks resemble blood vessels	
		Which tissue is the abnormal in angioid streaks?	
		Bruch's membrane	
Munson sign?	Yes	?Yes?	Yes



Q

Corneal Ectasias

	KCN	Keratoglobus	Pellucid Marginal Degeneration
How common		Keratoglobus has two strong syndromic associations--what are they?	
Age at onset		<ul style="list-style-type: none"> -Osteogenesis imperfecta (OI) -Ehlers-Danlos syndrome (EDS) 	
Thins where		What is the classic posterior-pole association for Ehlers-Danlos? and EDS? Angioid streaks	
Protrudes where		What is the classic mnemonic for angioid streaks? What is the DFE appearance of angioid streaks?	
Iron line present		What is the classic mnemonic for angioid streaks? What is the DFE appearance of angioid streaks?	
Spontaneous rupture	--	What is the classic mnemonic for angioid streaks? What is the DFE appearance of angioid streaks?	
Steepens where	--	What is the classic mnemonic for angioid streaks? What is the DFE appearance of angioid streaks?	
Munson sign	--	What is the classic mnemonic for angioid streaks? What is the DFE appearance of angioid streaks?	
		Yes	

A

Corneal Ectasias



	KCN	Keratoglobus	Pellucid Marginal Degeneration
How common		Keratoglobus has two strong syndromic associations--what are they?	
Age at onset		<ul style="list-style-type: none"> -Osteogenesis imperfecta (OI) -Ehlers-Danlos syndrome (EDS) 	
Thins where		What is the classic posterior-pole association for Ehlers-Danlos?	
Protrudes where		What is the classic mnemonic for angioid streaks?	
Iron line present		PEPSI	
Spontaneous rupture	--P		
Steepens where	--E		
Munson sign	--P		
	--S		
	--I		
			Yes

Angioid streaks

What is the classic mnemonic for angioid streaks?

PEPSI

--P

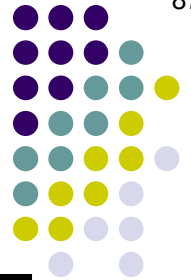
--E

--P

--S

--I

Yes



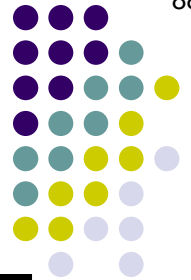
Q

Corneal Ectasias

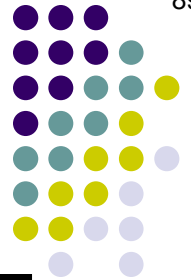
	KCN	Keratoglobus	Pellucid Marginal Degeneration
How common		Keratoglobus has two strong syndromic associations--what are they?	
Age at onset		<ul style="list-style-type: none"> -Osteogenesis imperfecta (OI) -Ehlers-Danlos syndrome (EDS) 	
Thins where		What is the classic posterior-pole association for Ehlers-Danlos?	
Protrudes where		Angioid streaks	
Iron line present		What is the classic mnemonic for angioid streaks?	
Spontaneous rupture		PEPSI	
Steepens where		What does each letter stand for?	
Munson sign		<ul style="list-style-type: none"> --P --Ehlers-Danlos --P --S --I 	
			Yes

A

Corneal Ectasias



	KCN	Keratoglobus	Pellucid Marginal Degeneration
How common		Keratoglobus has two strong syndromic associations--what are they?	
Age at onset		<ul style="list-style-type: none"> --Osteogenesis imperfecta (OI) --Ehlers-Danlos syndrome (EDS) 	
Thins where		What is the classic posterior-pole association for Ehlers-Danlos? Angioid streaks	
Protrudes where		What is the classic mnemonic for angioid streaks? PEPSI	
Iron line present		What does each letter stand for? -- P seudoxanthoma elasticum -- E hlers-Danlos -- P aget's dz of bone -- S ickle-cell dz -- I diopathic	
Spontaneous rupture			
Steepens where			
Munson sign		Yes	



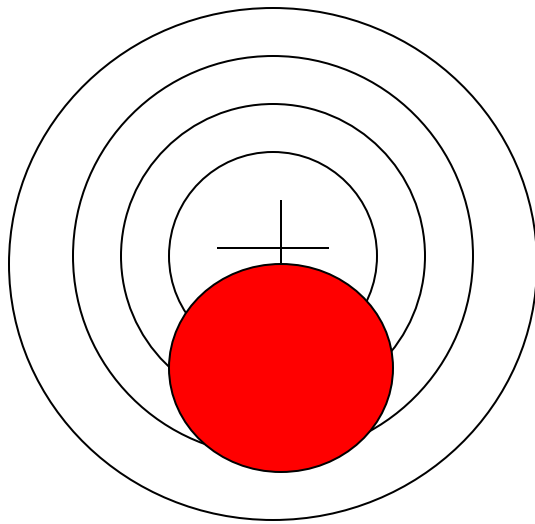
Corneal Ectasias

	KCN	Keratoglobus	Pellucid Marginal Degeneration
How common		Keratoglobus has two strong syndromic associations--what are they?	
Age at onset		<ul style="list-style-type: none"> --Osteogenesis imperfecta (OI) --Ehlers-Danlos syndrome (EDS) 	
Thins where		<p>For more on angioid streaks, see slide-set R61</p> <p>Angioid streaks</p>	
Protrudes where		<p>What is the classic mnemonic for angioid streaks?</p> <p>PEPSI</p>	
Iron line present		<p>What does each letter stand for?</p> <ul style="list-style-type: none"> --Pseudoxanthoma elasticum --Ehlers-Danlos --Paget's dz of bone --Sickle-cell dz --Idiopathic 	
Spontaneous rupture			
Steepens where			
Munson sign			
		Yes	

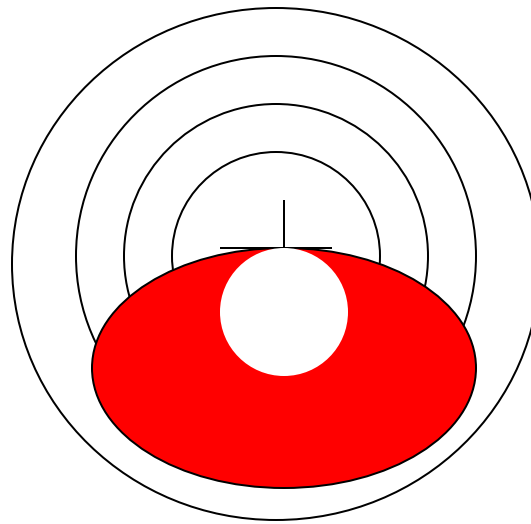


Q

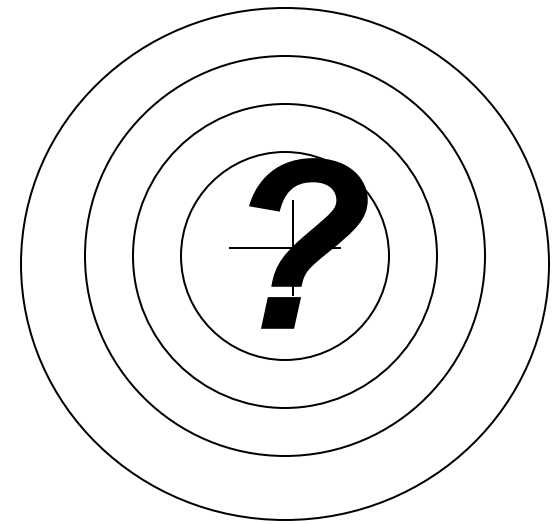
Corneal Ectasias



Condition?



Condition?



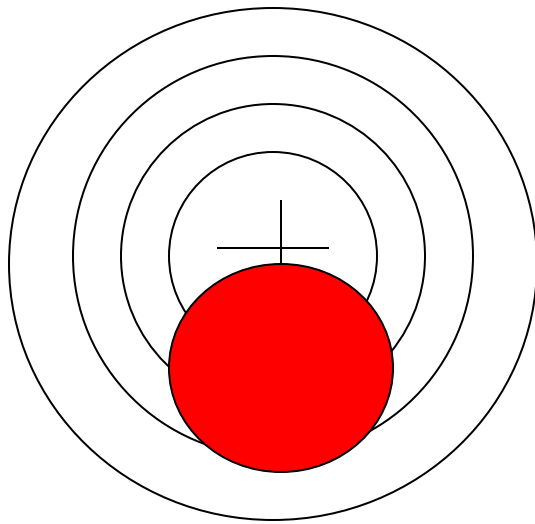
Condition?

These drawings represent typical corneal topography maps for keratoconus, keratoglobus and pellucid marginal degeneration. (The '?' map indicates this condition's topography has yet to be well described.) Which is which?

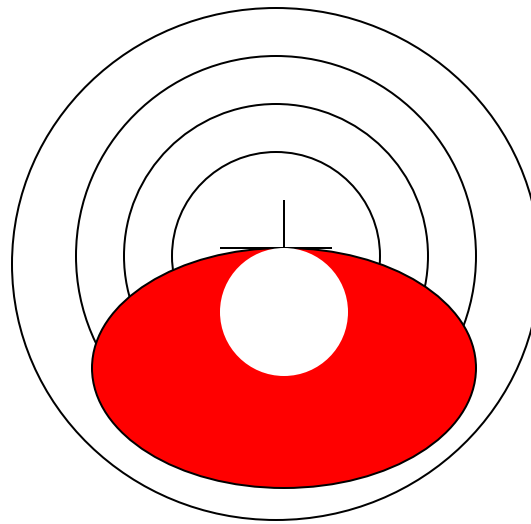


A

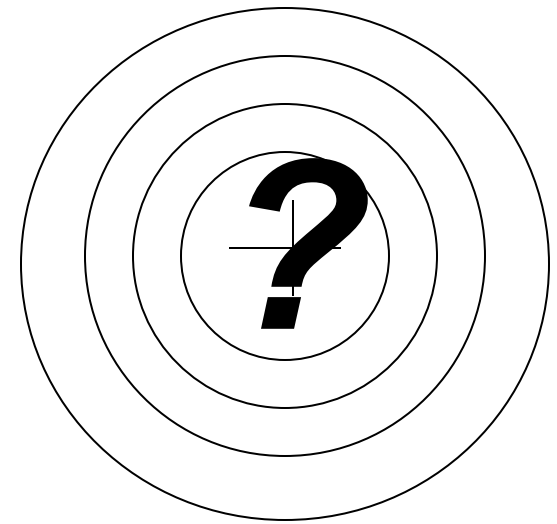
Corneal Ectasias



Keratoconus

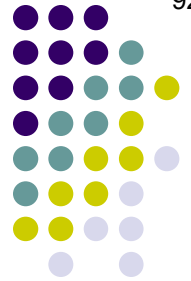


Pellucid marginal degeneration



Keratoglobus

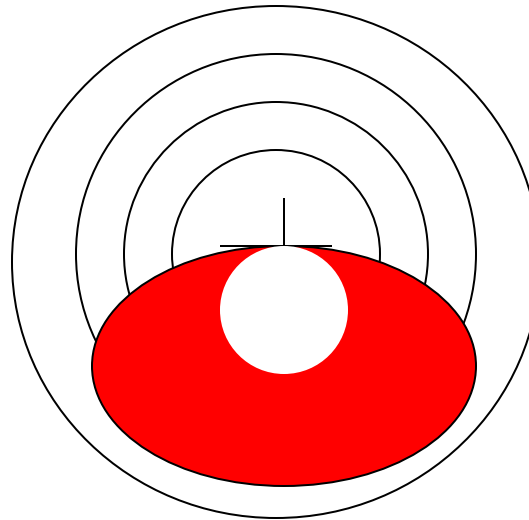
These drawings represent typical corneal topography maps for keratoconus, keratoglobus and pellucid marginal degeneration. (The '?' map indicates this condition's topography has yet to be well described.) Which is which?



Q

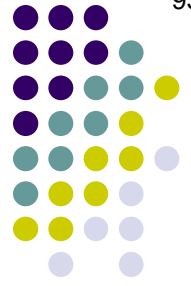
Corneal Ectasias

‘Lobster claw’



Pellucid marginal degeneration

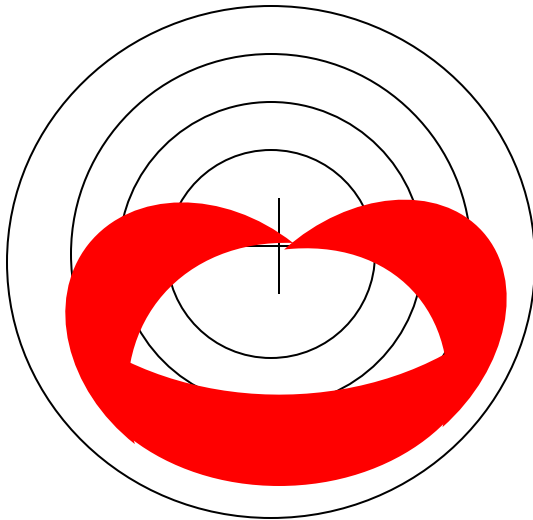
This PMD topography pattern is called the ‘lobster claw.’
What other, similar maps are also seen in PMD?



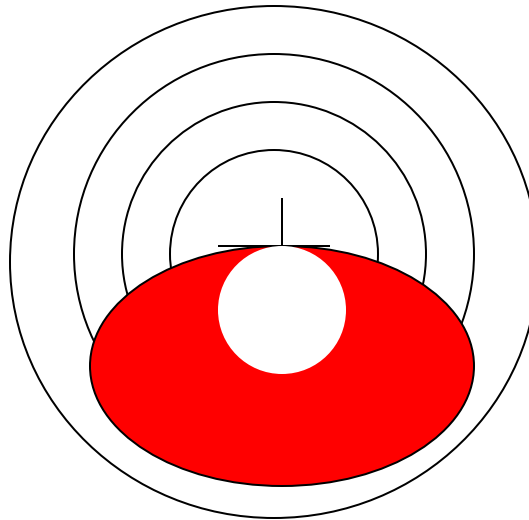
A

Corneal Ectasias

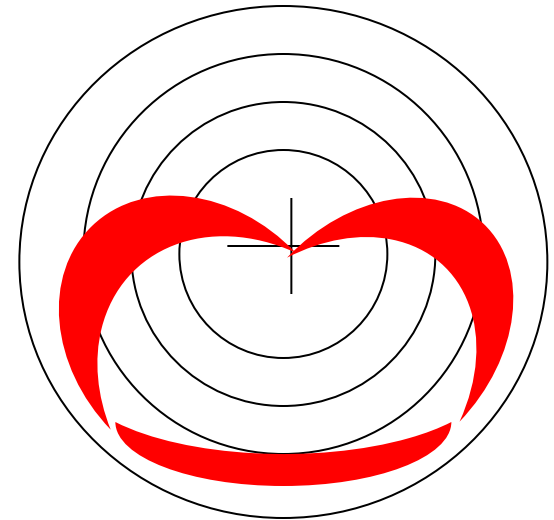
'Kissing doves'



'Lobster claw'



'The moustache'



**Pellucid marginal degeneration
(All 3)**

This PMD topography pattern is called the 'lobster claw.'
What other, similar maps are also seen in PMD?

Other terms used to describe topography in PMD: 'The butterfly'; 'the teardrop'

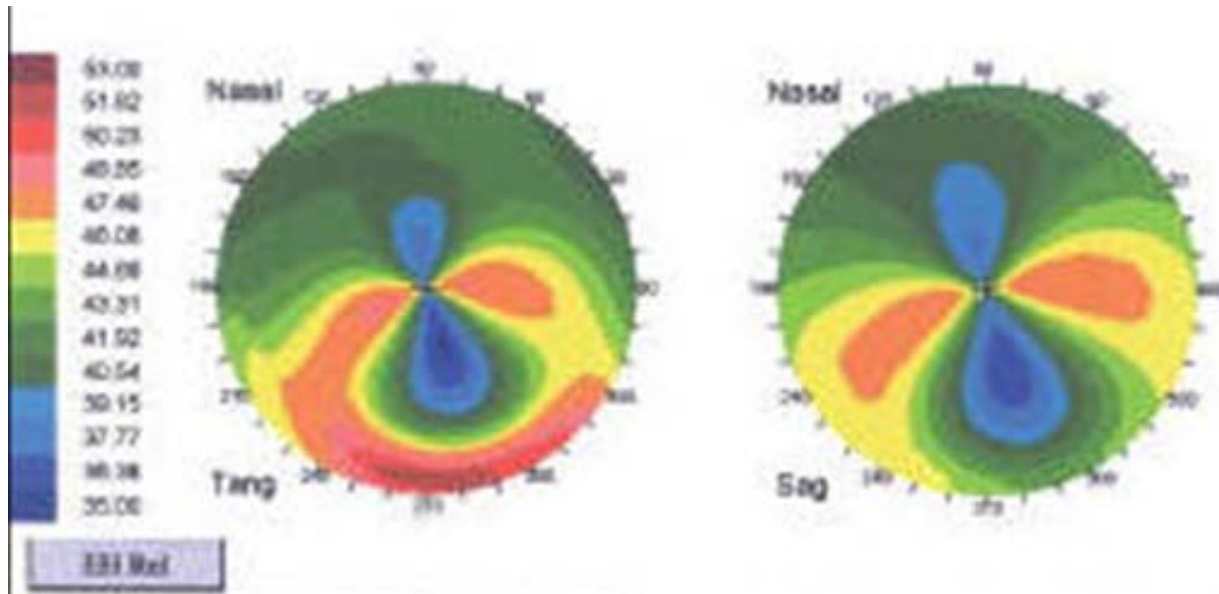
Corneal Ectasias



‘Kissing doves’

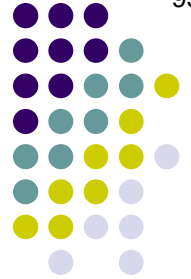
‘Lobster claw’

‘The moustache’



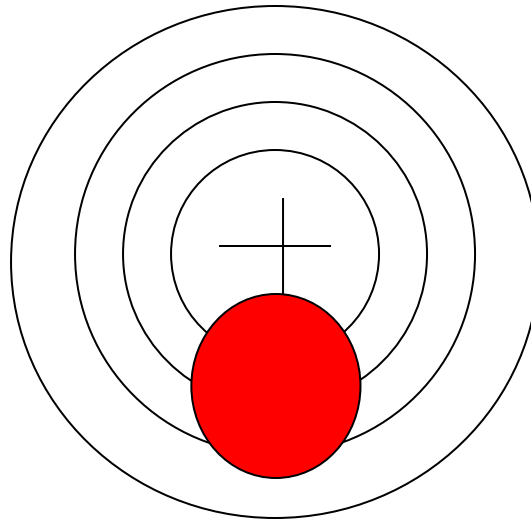
Pellucid marginal degeneration

No question—proceed when ready



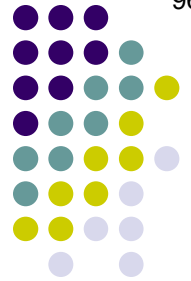
Q

Corneal Ectasias



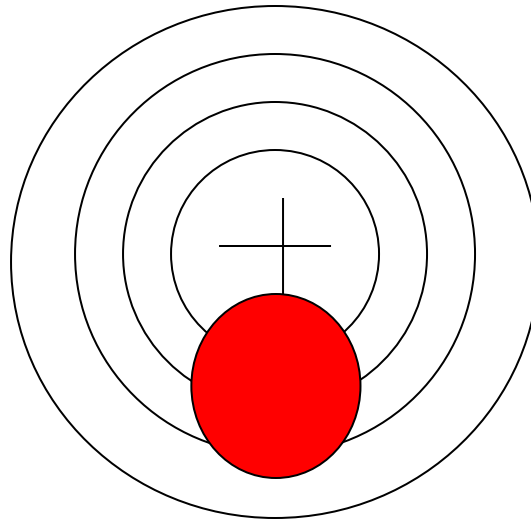
Topography in keratoconus reveals

three words



A

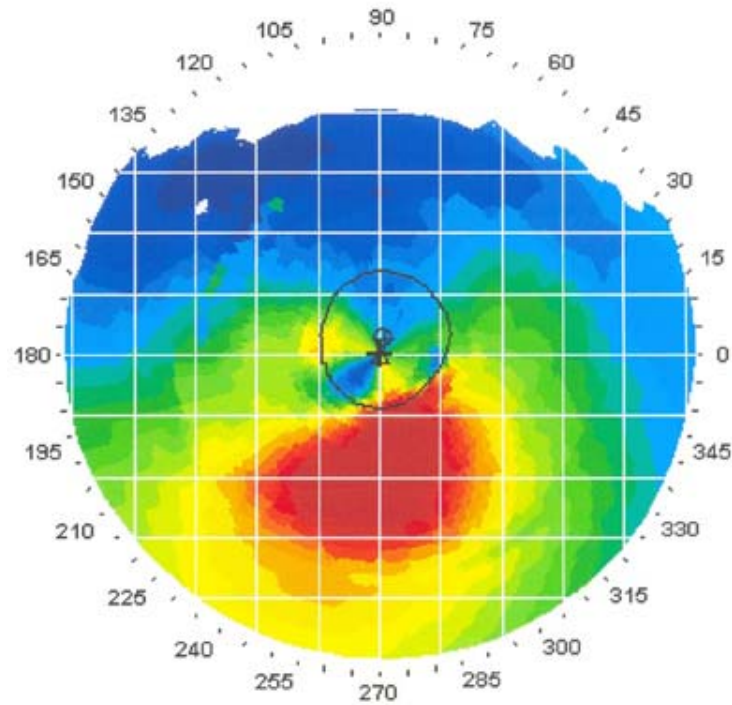
Corneal Ectasias



Topography in keratoconus reveals
inferior corneal steepening

A

Corneal Ectasias

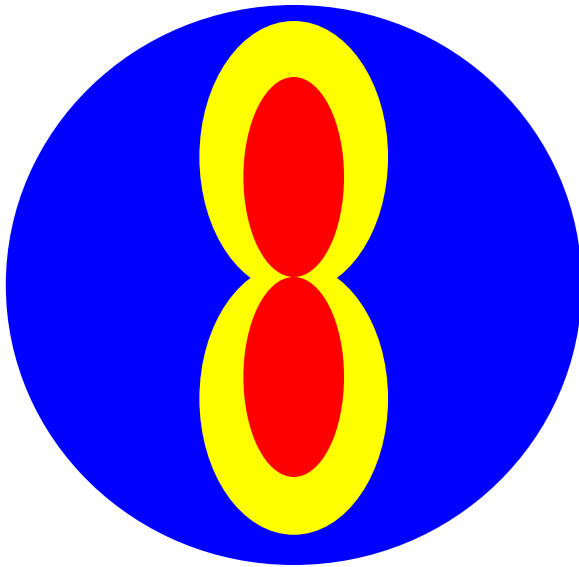
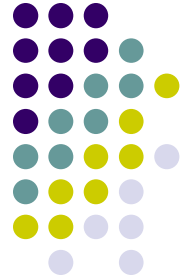


Topography in keratoconus reveals
inferior corneal steepening

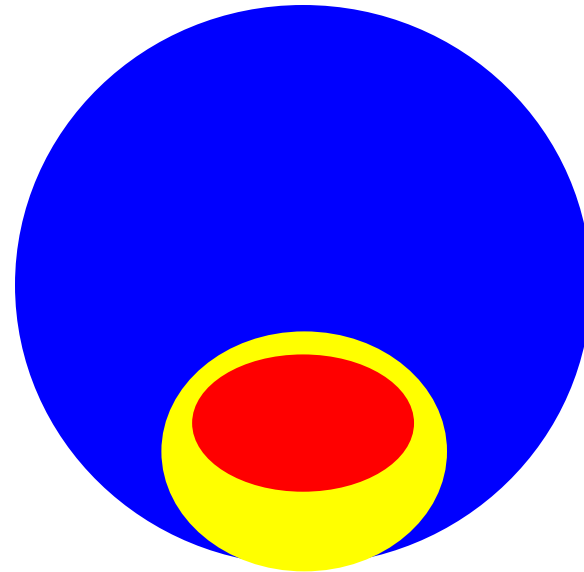
No question—proceed when ready

Q

Corneal Ectasias



?

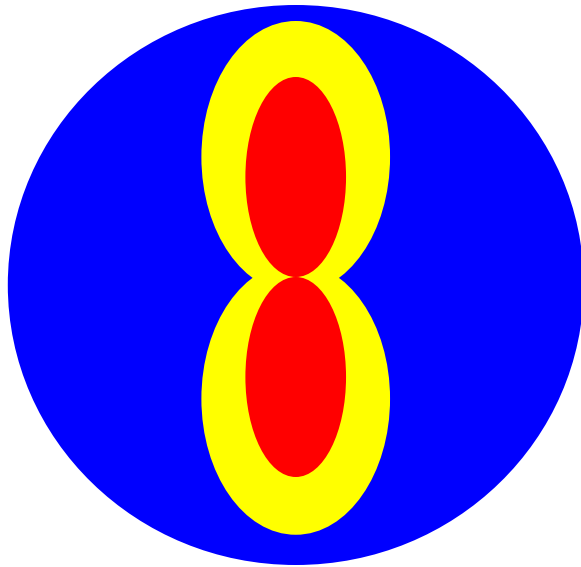
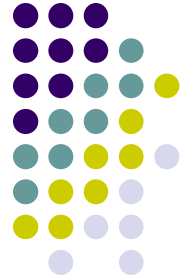


?

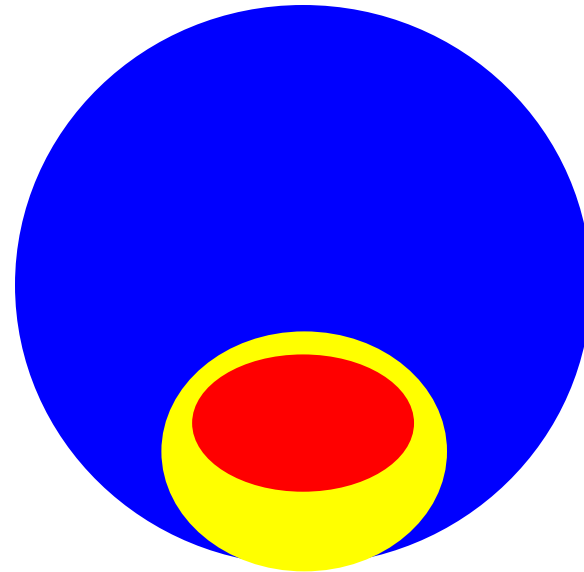
What are these?

A

Corneal Ectasias



With-the-rule astigmatism

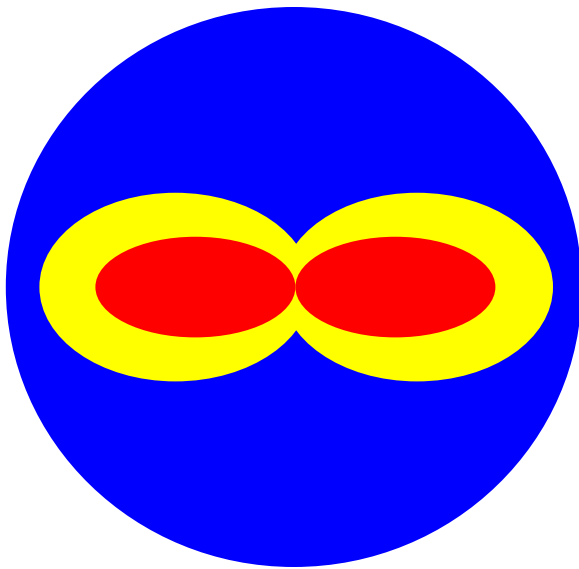
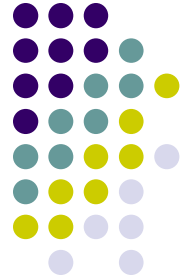


KCN

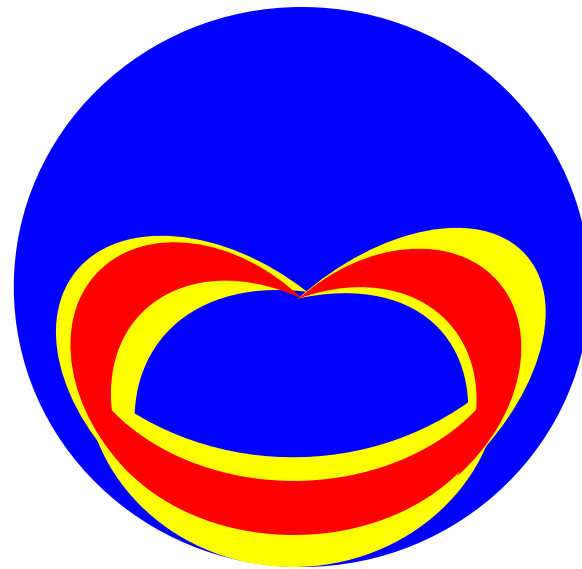
What are these?

Q

Corneal Ectasias



?

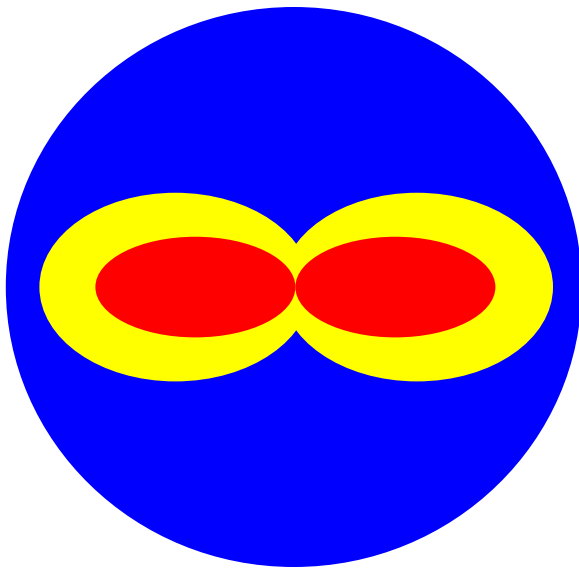
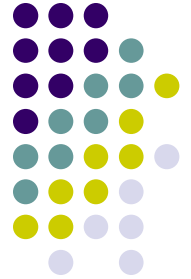


?

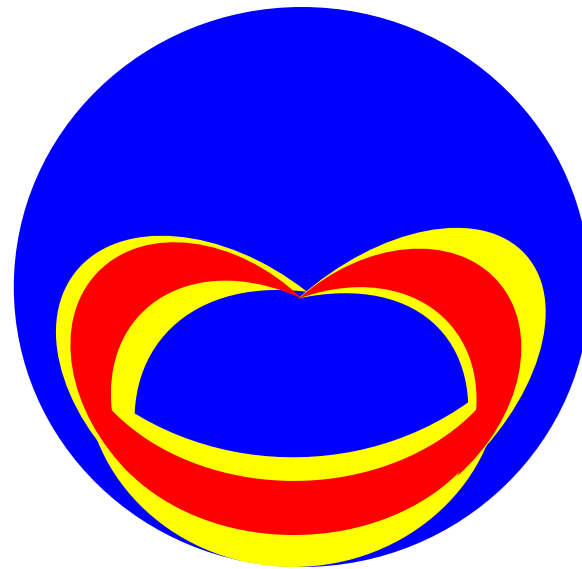
What are these?

A

Corneal Ectasias



Against-the-rule astigmatism

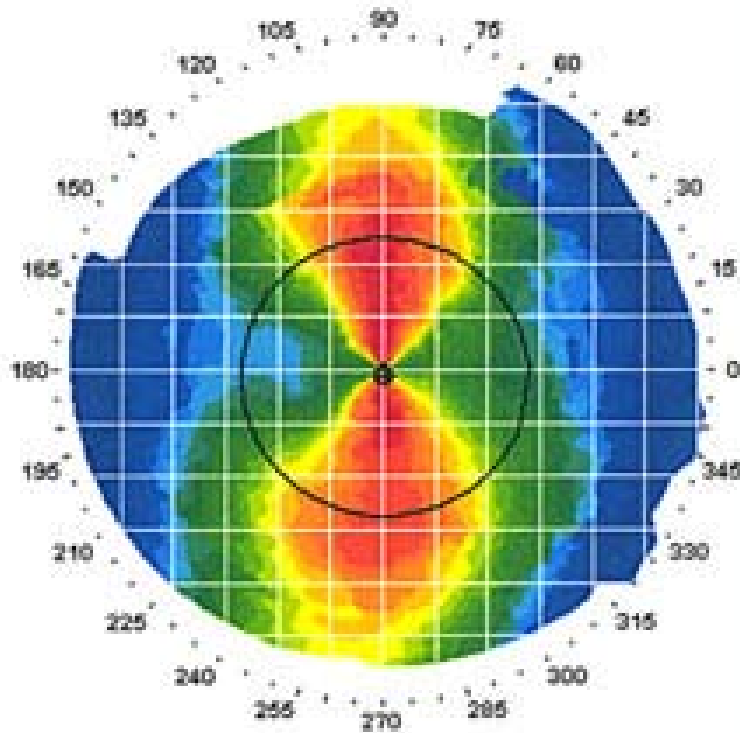


Pellucid

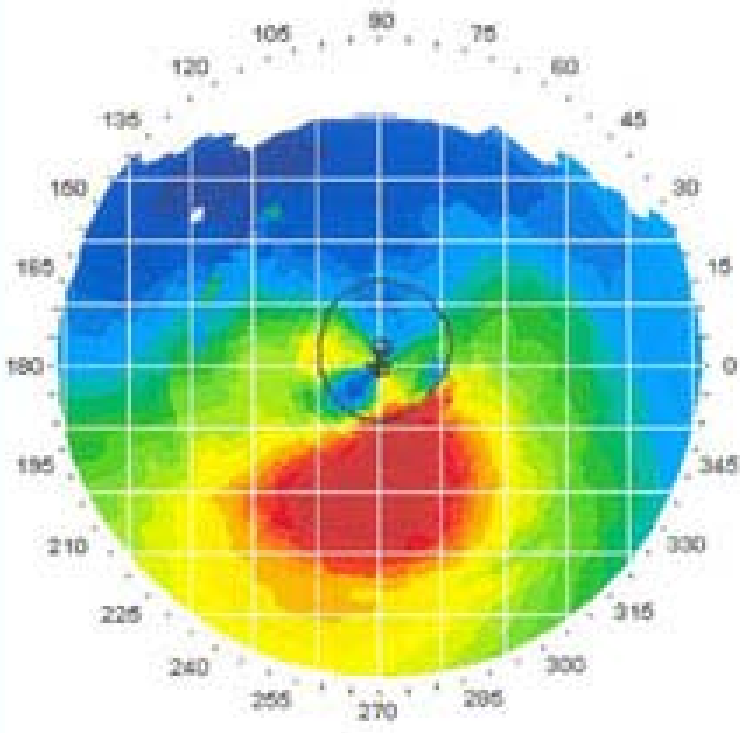
What are these?

Q

Corneal Ectasias



?

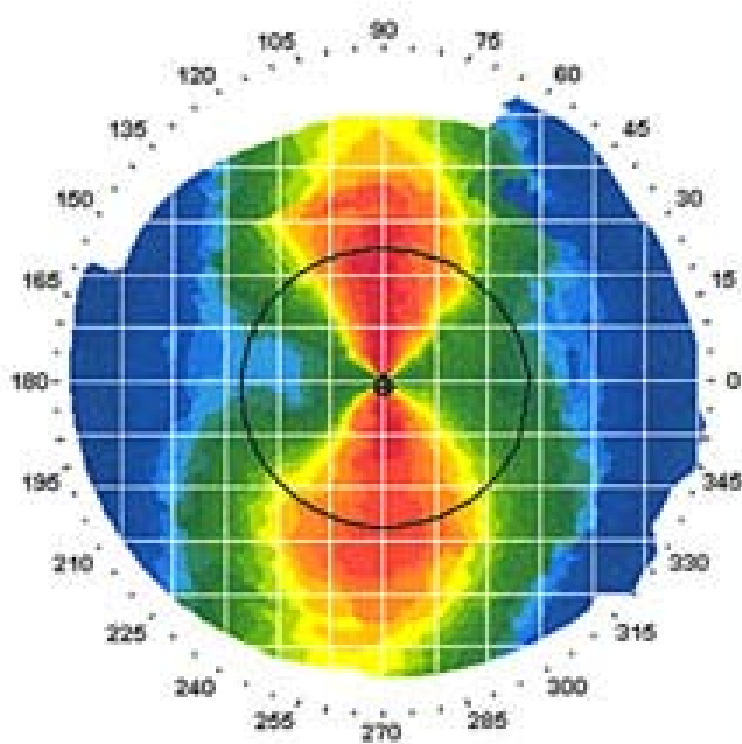


?

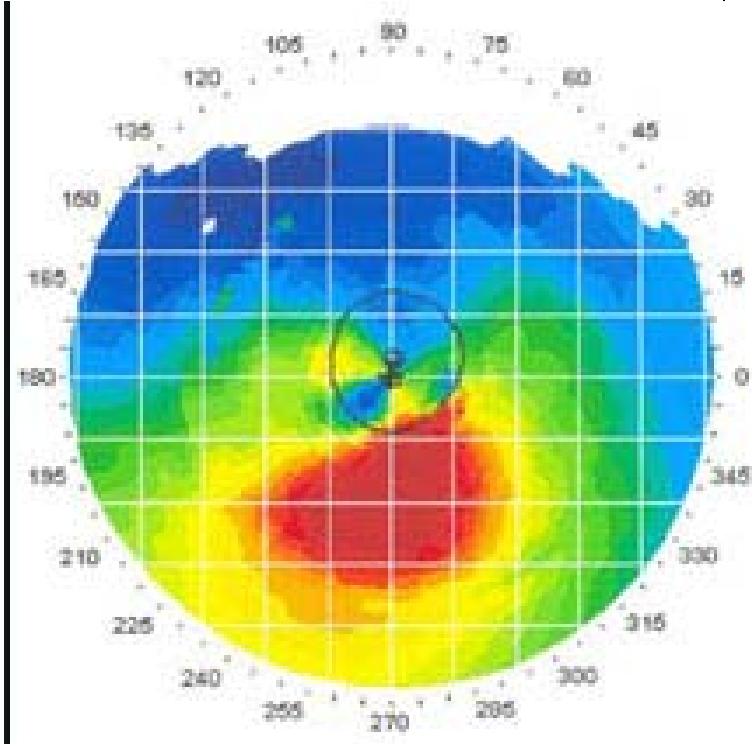
What are these?

A

Corneal Ectasias



With-the-rule astigmatism

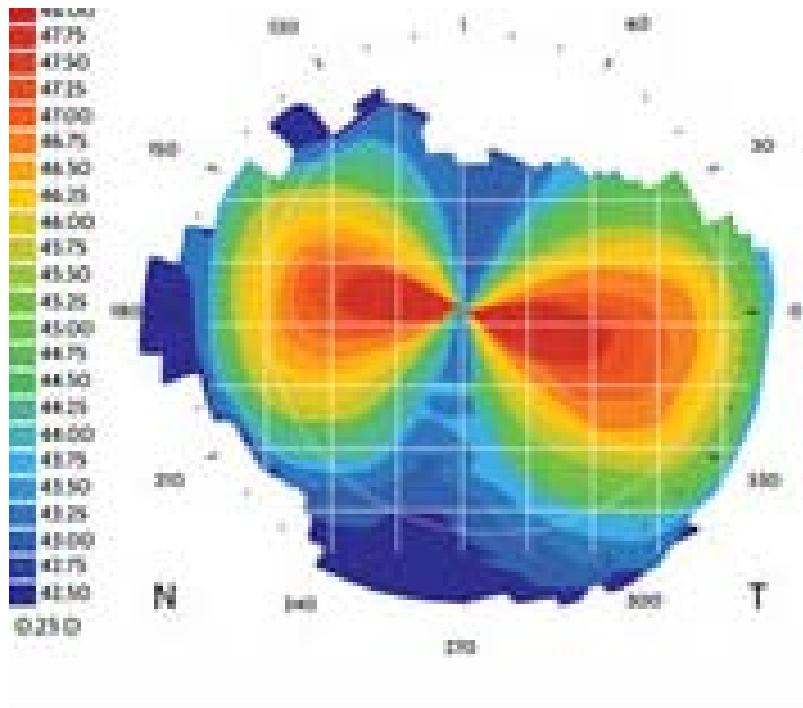
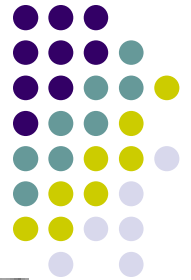


KCN

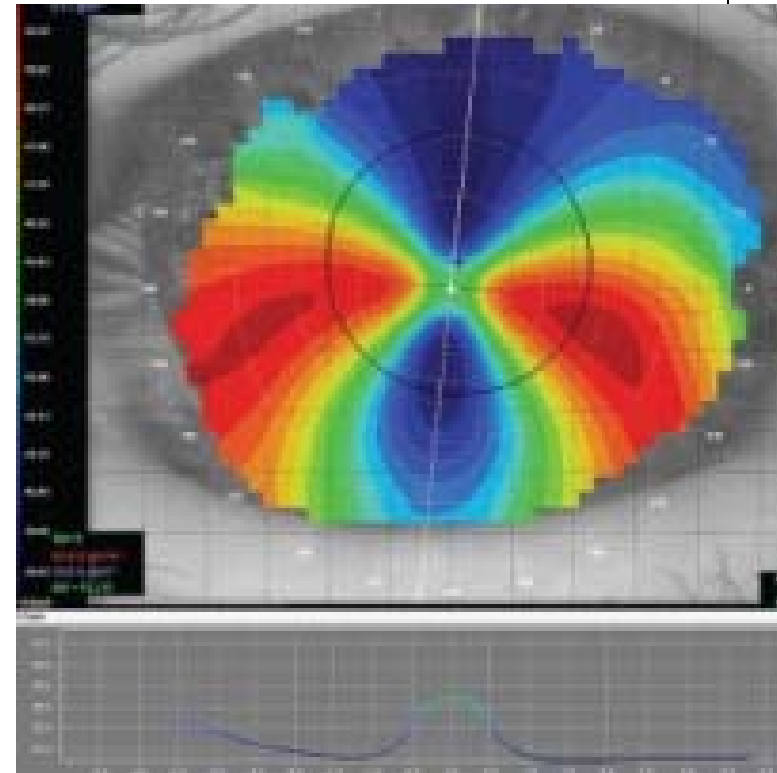
What are these?

Q

Corneal Ectasias



?

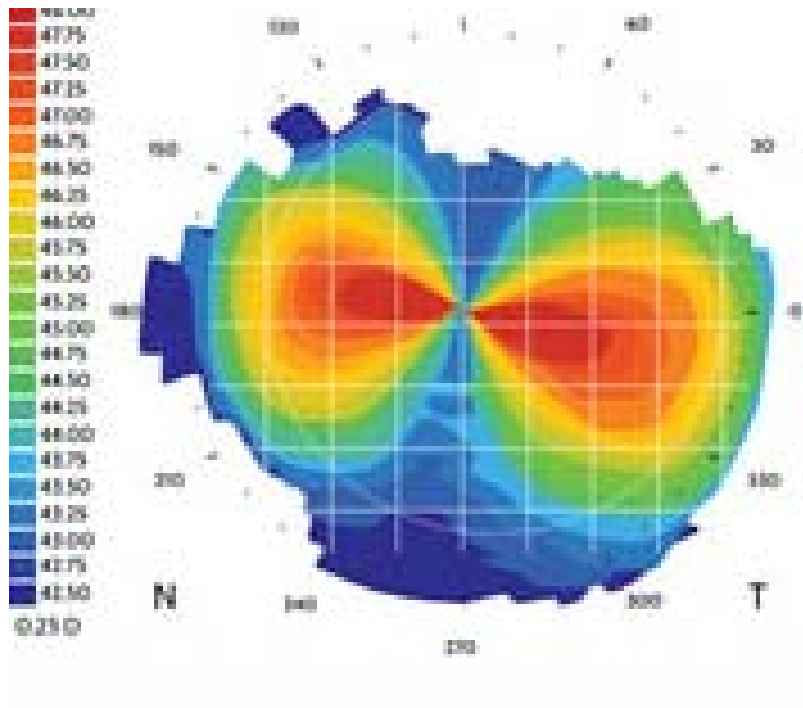


?

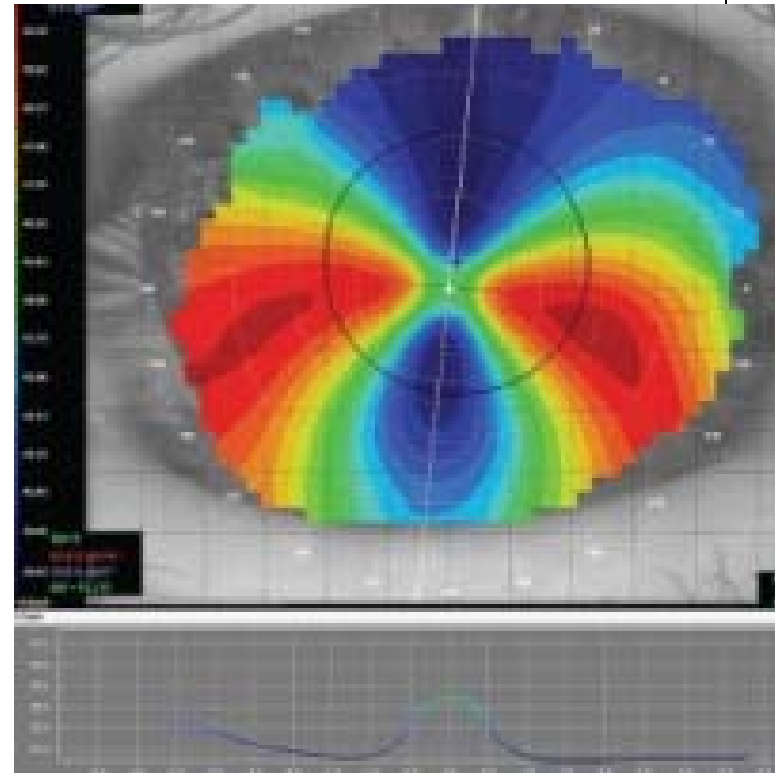
What are these?

A

Corneal Ectasias



Against-the-rule astigmatism

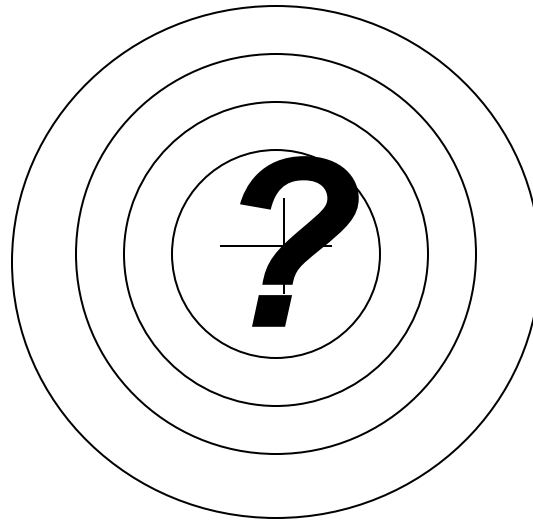


Pellucid

What are these?

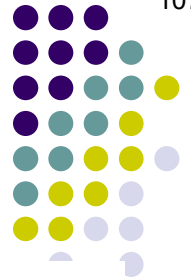


Corneal Ectasias



Corneal topography in keratoglobus has not been well delineated, and thus I would not expect to see such a map on the OKAP and/or Boards!

No question—proceed when ready



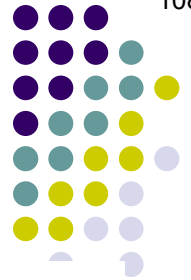
Q

Corneal Ectasias

- What are the 7 classic associations of keratoconus?



Mnemonic is...



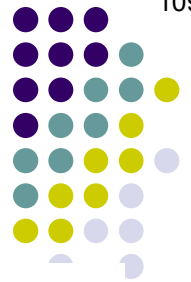
Q/A

Corneal Ectasias

- What are the 7 classic associations of keratoconus?

- F
- L
- A
- M
- E
- D

*Mnemonic is...**FLAMED***



A

Corneal Ectasias

- What are the 7 classic associations of keratoconus?
 - **F**loppy eyelid syndrome
 - **L**eber's congenital amaurosis
 - **A**topic disease (including **A**KC)
 - **M**itral valve prolapse
 - **E**hlers-Danlos
 - **D**own syndrome

*Mnemonic is...**FLAMED***