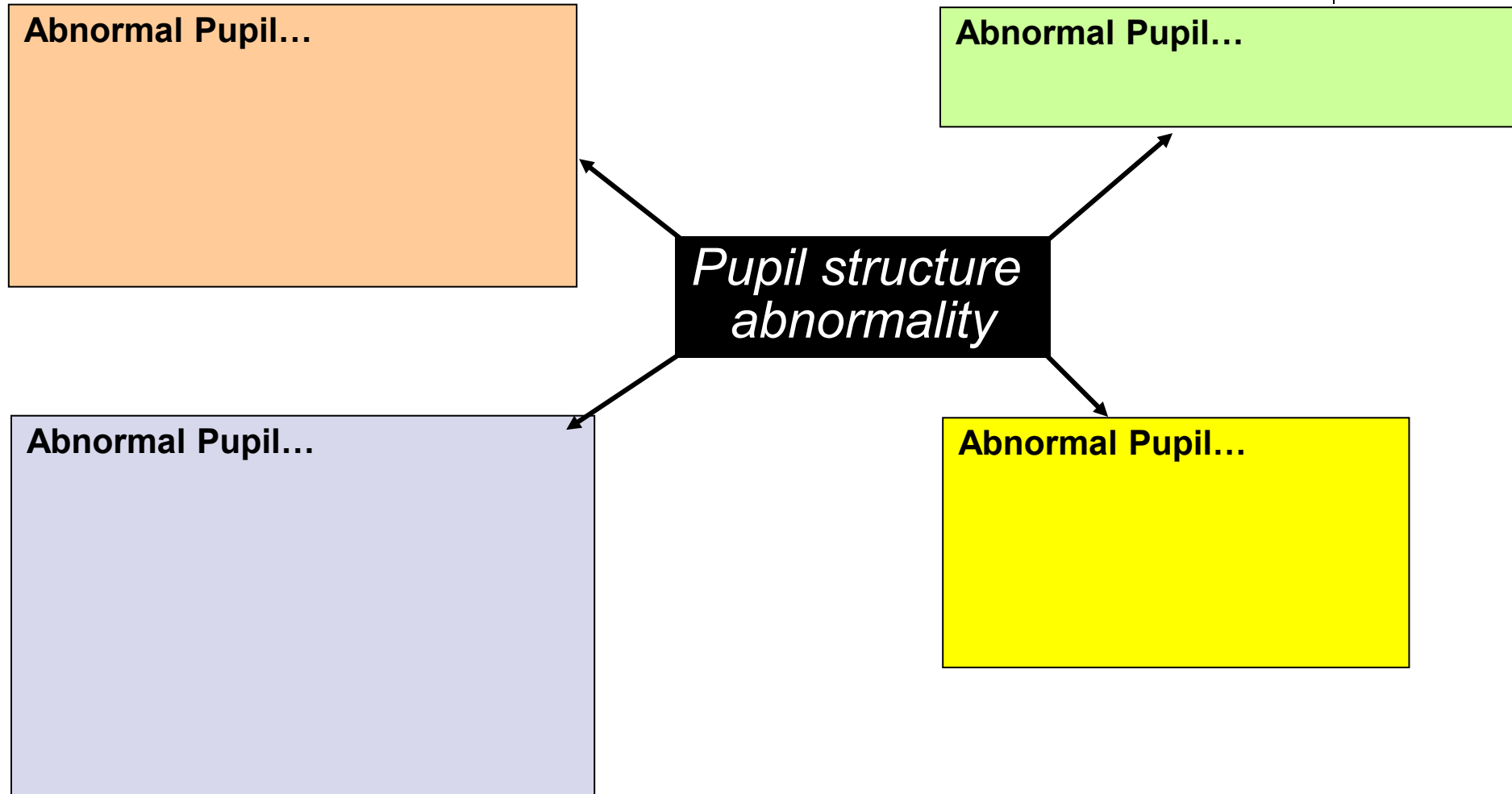
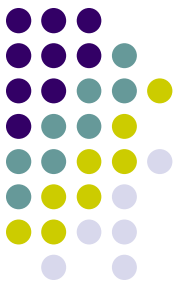
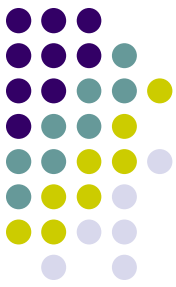


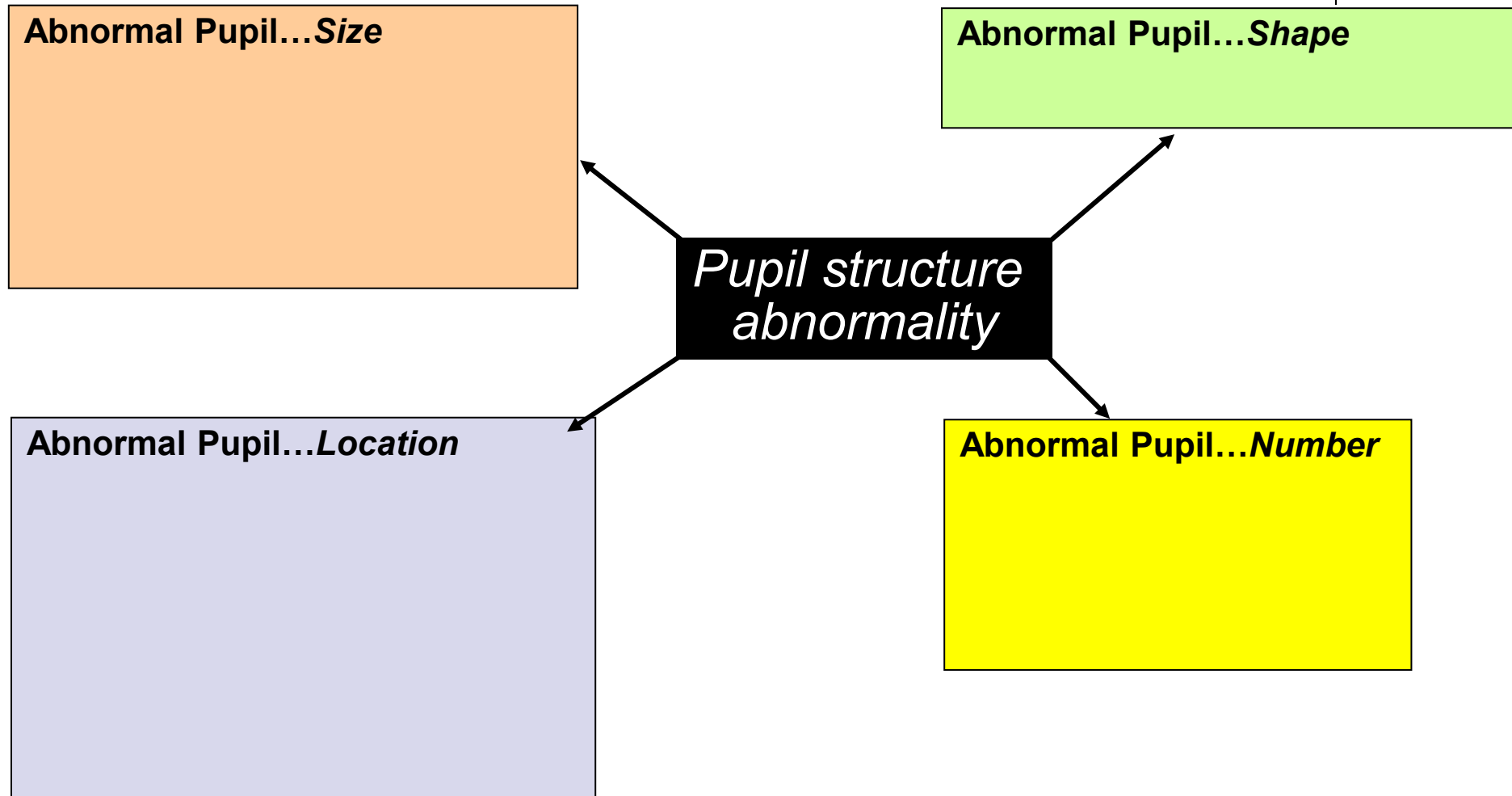
Abnormalities of Pupil Structure

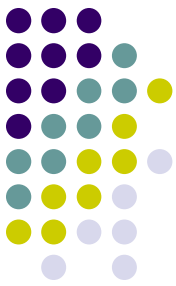
(Other than aniridia)



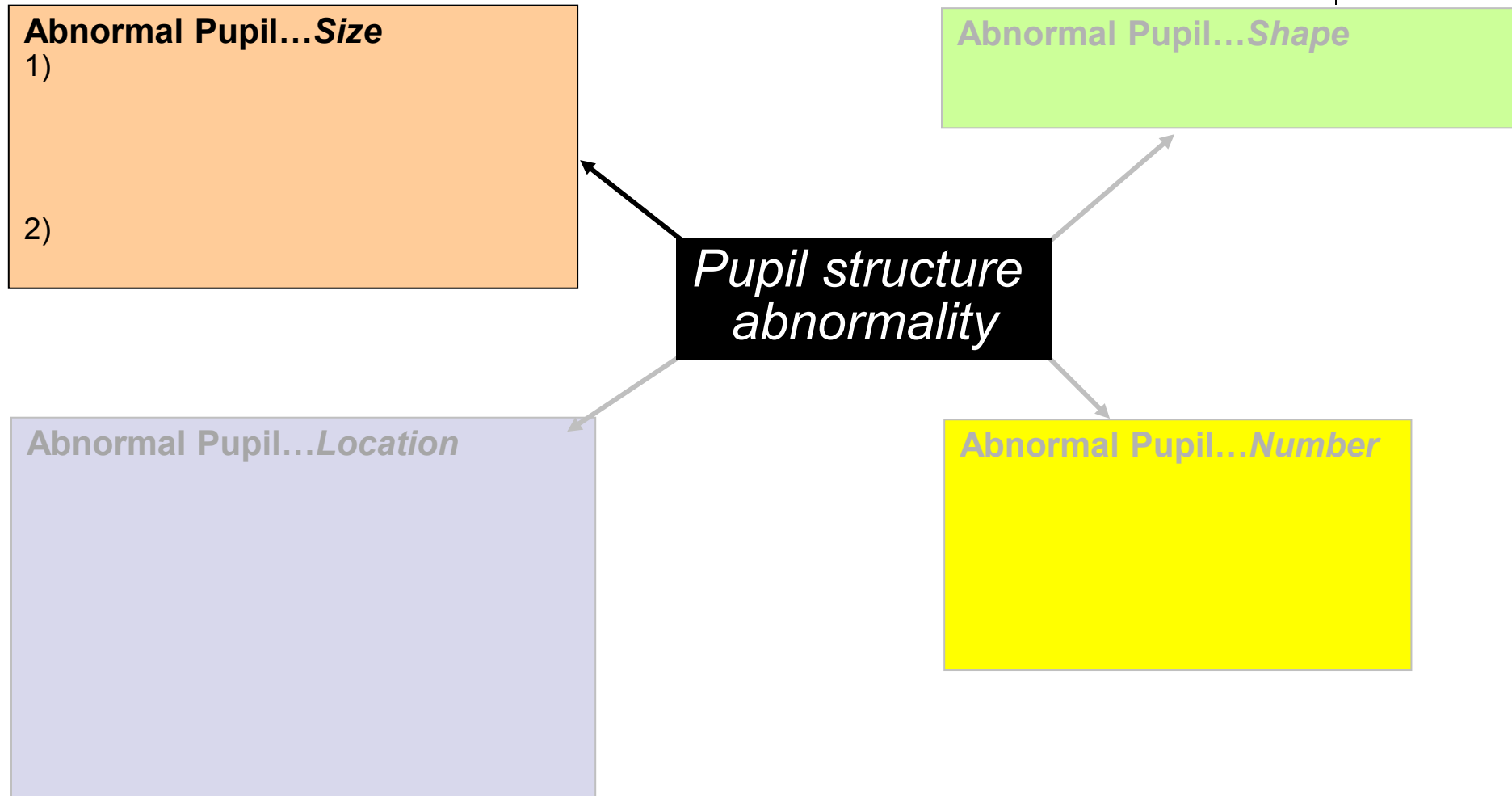


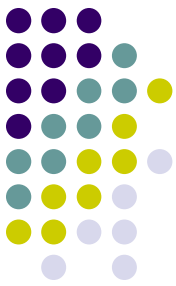
Abnormalities of Pupil Structure





Abnormalities of Pupil Structure





Abnormalities of Pupil Structure

Abnormal Pupil...Size

1) Congenital



2) Congenital

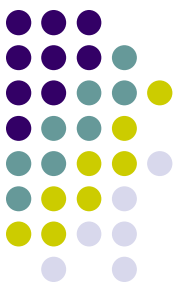


Abnormal Pupil...Shape

*Pupil structure
abnormality*

Abnormal Pupil...Location

Abnormal Pupil...Number



Abnormalities of Pupil Structure

Abnormal Pupil...Size

1) Congenital **mios**is (*microcoria*)

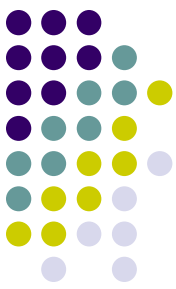
2) Congenital **mydri**asis

Abnormal Pupil...Shape

*Pupil structure
abnormality*

Abnormal Pupil...Location

Abnormal Pupil...Number



Abnormalities of Pupil Structure

Abnormal Pupil...Size

- 1) Congenital **miosis** (*microcoria*)
--Due to dilator absence/malformation,
or fibrous contraction

- 2) Congenital **mydriasis**

Abnormal Pupil...Shape

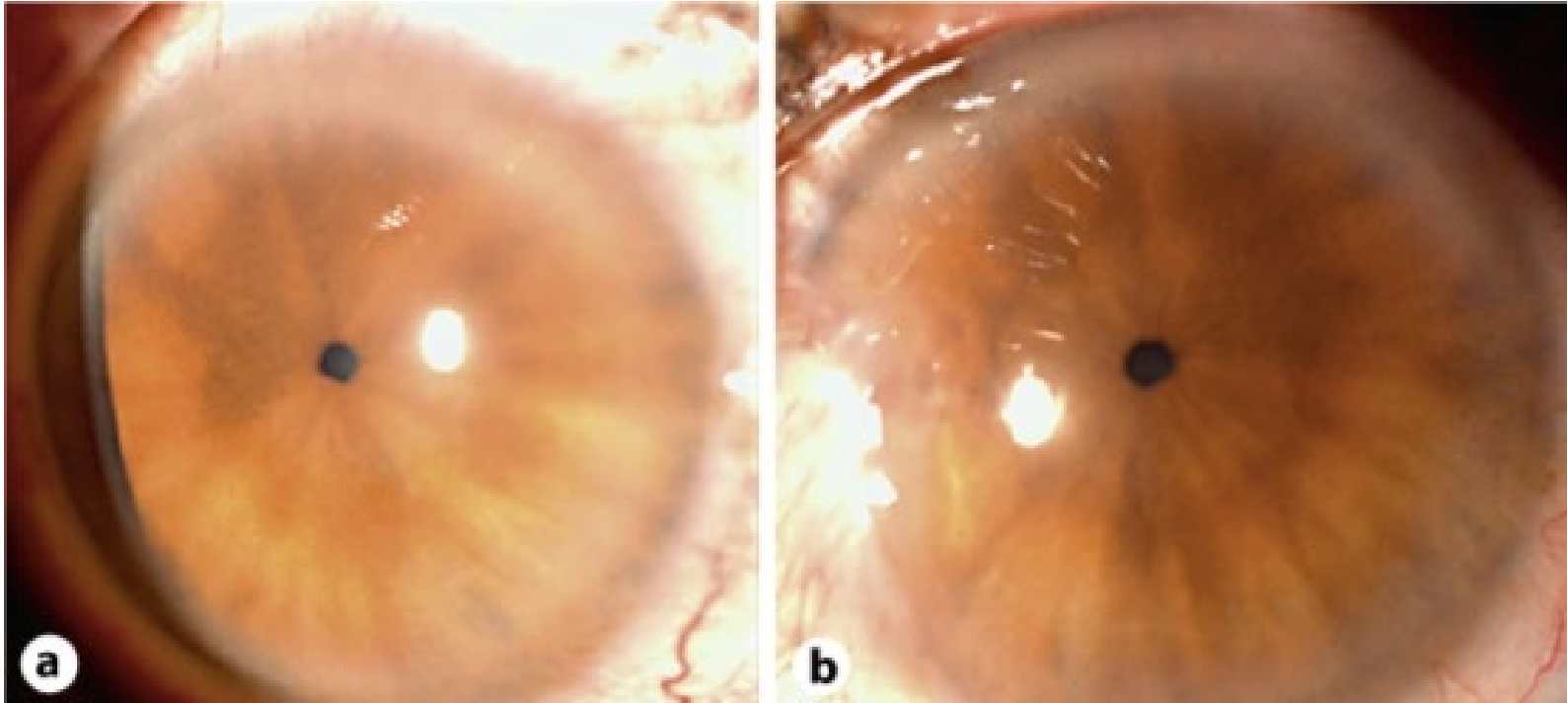
Pupil structure abnormality

Abnormal Pupil...Location

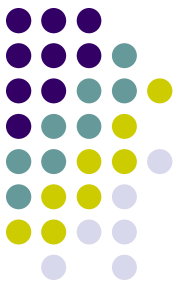
Abnormal Pupil...Number



Abnormalities of Pupil Structure



Congenital microcoria



Abnormalities of Pupil Structure

Abnormal Pupil...Size

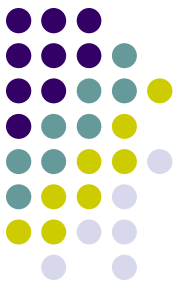
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Abnormal Pupil...Shape

Pupil structure abnormality

Abnormal Pupil...Location

Abnormal Pupil...Number



Abnormalities of Pupil Structure

Abnormal Pupil...Size

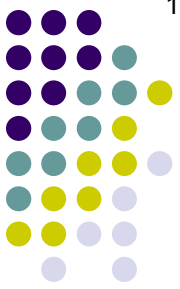
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Abnormal Pupil...Shape

Pupil structure abnormality

Abnormal Pupil...Location

Abnormal Pupil...Number



Abnormalities of Pupil Structure

Abnormal Pupil...Size

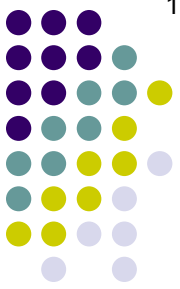
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 - Probably part of the aniridia spectrum

Abnormal Pupil...Shape

Pupil structure abnormality

Abnormal Pupil...Location

Abnormal Pupil...Number

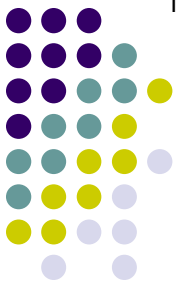


Abnormalities of Pupil Structure



The patient was a 73-year-old female with bilateral congenital mydriasis and no other abnormalities. The pupils reacted almost imperceptibly to light.

Congenital microcoria



Abnormalities of Pupil Structure

Abnormal Pupil...Size

- 1) Congenital **mios**is (*microcoria*)
 - Due to dilator absence/malformation, or fibrous contraction
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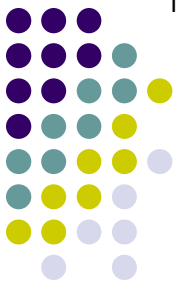
Abnormal Pupil...Shape

--**Dyscoria** =

Pupil structure abnormality

Abnormal Pupil...Location

Abnormal Pupil...Number



Abnormalities of Pupil Structure

Abnormal Pupil...Size

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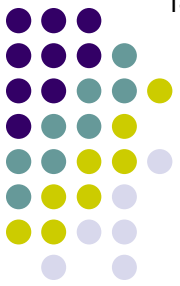
Abnormal Pupil...Shape

--**Dyscoria** = congenital misshapen pupil
 --e.g.,

Pupil structure abnormality

Abnormal Pupil...Location

Abnormal Pupil...Number



Abnormalities of Pupil Structure

Abnormal Pupil...Size

- 1) Congenital **mios**is (*microcoria*)
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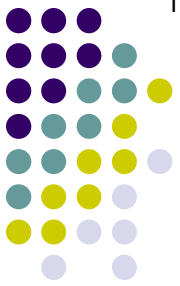
Abnormal Pupil...Shape

--**Dyscoria** = congenital misshapen pupil
--e.g., coloboma

Pupil structure abnormality

Abnormal Pupil...Location

Abnormal Pupil...Number



Abnormalities of Pupil Structure



Slit-shaped dyscoric pupil OS (note also the ipsilateral microcornea)

Dyscoria



Abnormalities of Pupil Structure

Abnormal Pupil...Size

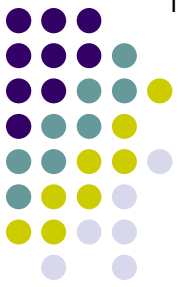
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Abnormal Pupil...Shape

- Dyscoria** = congenital misshapen pupil
- e.g., **coloboma**

Pupil structure abnormality

Abn		Location			
	Typical iris coloboma	?			
	Atypical iris coloboma	?			



Abnormalities of Pupil Structure

Abnormal Pupil...Size

- 1) Congenital **miosis** (*microcoria*)
 - Due to dilator absence/malformation, or fibrous contraction
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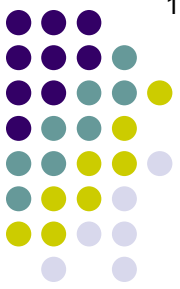
Abnormal Pupil...Shape

- Dyscoria** = congenital misshapen pupil
- e.g., **coloboma**

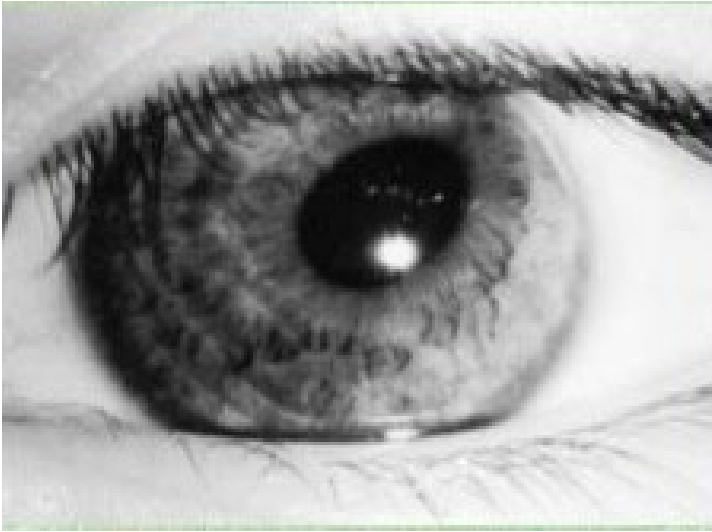
Pupil structure abnormality

Abn

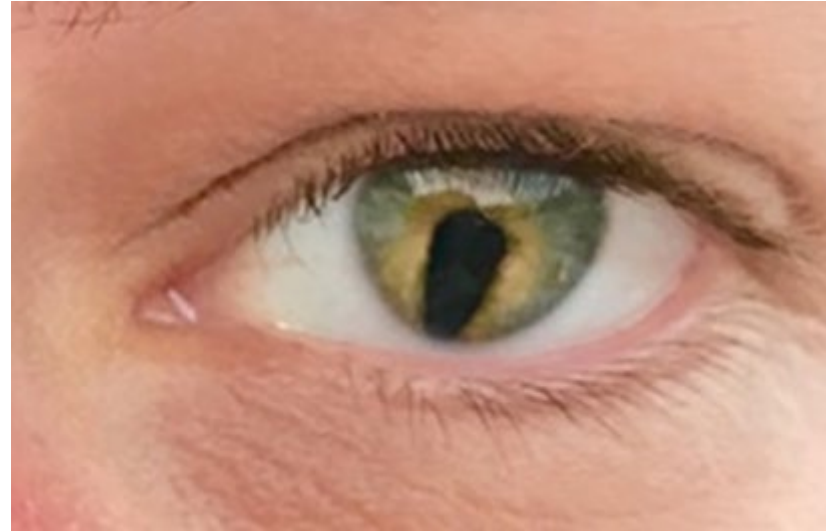
	Location			
Typical iris coloboma	Inferonasal			
Atypical iris coloboma	Anywhere <i>but</i> inferonasal			



Abnormalities of Pupil Structure

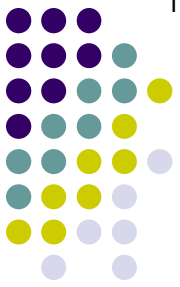


Atypical



Typical

Coloboma



Abnormalities of Pupil Structure

Abnormal Pupil...Size

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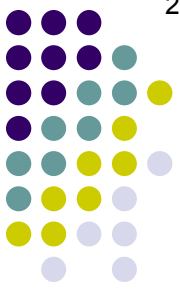
Abnormal Pupil...Shape

- Dyscoria** = congenital misshapen pupil
- e.g., **coloboma**

Pupil structure abnormality

Abn

	<i>Location</i>	<i>Etiology</i>		
Typical iris coloboma	Inferonasal	?		
Atypical iris coloboma	Anywhere <i>but</i> inferonasal	?		



Abnormalities of Pupil Structure

Abnormal Pupil...Size

- 1) Congenital **miosis** (*microcoria*)
 - Due to dilator absence/malformation, or fibrous contraction
 - Associated with multiple congenital ocular abnormalities
- 2) Congenital **mydriasis**
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Abnormal Pupil...Shape

- Dyscoria** = congenital misshapen pupil
- e.g., **coloboma**

Pupil structure abnormality

Abn

	<i>Location</i>	<i>Etiology</i>		
Typical iris coloboma	Inferonasal	Incomplete closure of embryonic fissure		
Atypical iris coloboma	Anywhere <i>but</i> inferonasal	Multiple etiologies implicated		



Abnormalities of Pupil Structure

Abnormal Pupil...Size

- 1) Congenital **miosis** (*microcoria*)
 - Due to dilator absence/malformation, or fibrous contraction
 - Associated with multiple congenital ocular abnormalities
- 2) Congenital **mydriasis**
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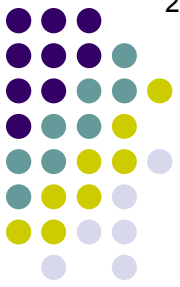
Abnormal Pupil...Shape

- Dyscoria** = congenital misshapen pupil
- e.g., **coloboma**

Pupil structure abnormality

Abr

	<i>Location</i>	<i>Etiology</i>	<i>Associated abnormalities</i>	
Typical iris coloboma	Inferonasal	Incomplete closure of embryonic fissure	?	
Atypical iris coloboma	Anywhere <i>but</i> inferonasal	Multiple etiologies implicated	?	



Abnormalities of Pupil Structure

Abnormal Pupil...Size

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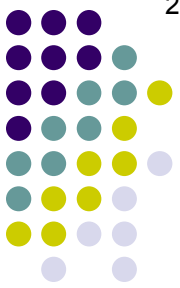
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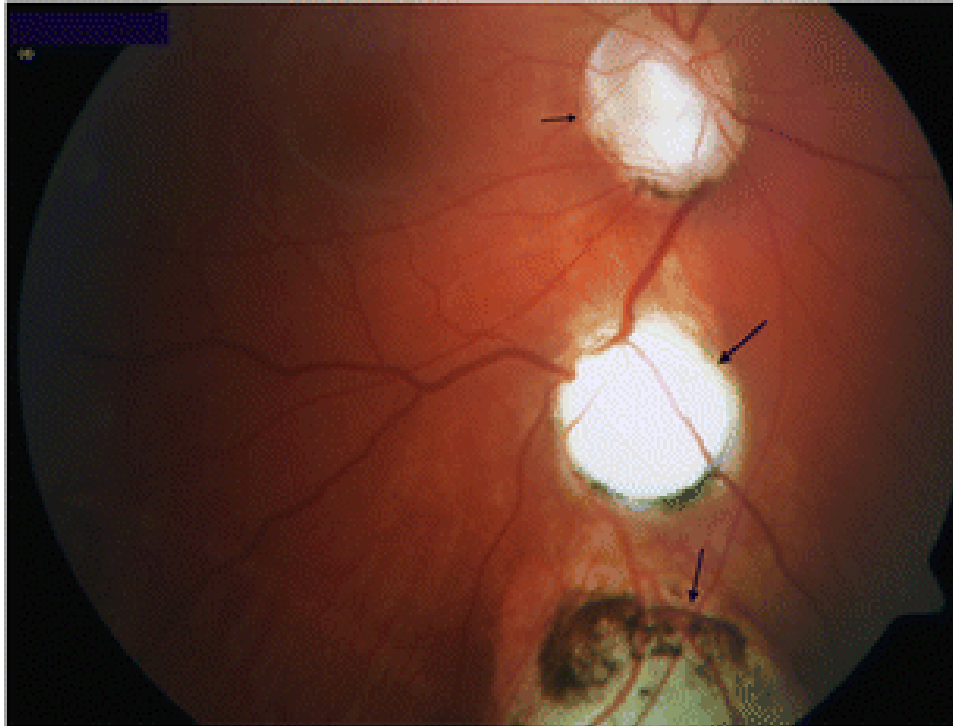
Pupil structure abnormality

Abn

	<i>Location</i>	<i>Etiology</i>	<i>Associated abnormalities</i>	
Typical iris coloboma	Inferonasal	Incomplete closure of embryonic fissure	Colobomas of CB, choroid, optic nerve, retina; nystagmus	
Atypical iris coloboma	Anywhere <i>but</i> inferonasal	Multiple etiologies implicated	None	

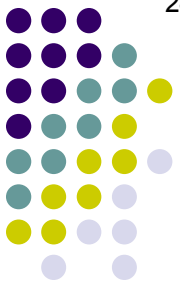


Abnormalities of Pupil Structure



Right eye illustrating optic disc coloboma (*small arrow*) and two retinochoroidal colobomas (*large arrows*)

Coloboma



Abnormalities of Pupil Structure

Abnormal Pupil...Size

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 - Due to dilator absence/malformation, or fibrous contraction
 - Associated with multiple congenital ocular abnormalities
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Abnormal Pupil...Shape

- Dyscoria** = congenital misshapen pupil
- e.g., **coloboma**

Pupil structure abnormality

Abn

	Location	Etiology	Associated abnormalities	Workup
Typical iris coloboma	Inferonasal	Incomplete closure of embryonic fissure	Colobomas of CB, choroid, optic nerve, retina; nystagmus	?
Atypical iris coloboma	Anywhere <i>but</i> inferonasal	Multiple etiologies implicated	None	?



Abnormalities of Pupil Structure

Abnormal Pupil...Size

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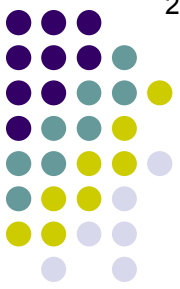
Abnormal Pupil...Shape

- Dyscoria** = congenital misshapen pupil
- e.g., **coloboma**

Pupil structure abnormality

Abn

	Location	Etiology	Associated abnormalities	Workup
Typical iris coloboma	Inferonasal	Incomplete closure of embryonic fissure	Colobomas of CB, choroid, optic nerve, retina; nystagmus	If 1+ other organ system abnormal: Do karyotype analysis with extended banding
Atypical iris coloboma	Anywhere <i>but</i> inferonasal	Multiple etiologies implicated	None	None



Abnormalities of Pupil Structure

Abnormal Pupil...Size

- 1) Congenital **mios**is (*microcoria*)
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Abnormal Pupil...Shape

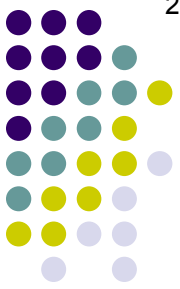
--**Dyscoria** = congenital misshapen pupil
 --e.g., coloboma

Pupil structure abnormality

Abnormal Pupil...Location

Abnormal Pupil...Number

--**Polycoria** =



Abnormalities of Pupil Structure

Abnormal Pupil...Size

- 1) Congenital **mios**is (*microcoria*)
 - Due to dilator absence/malformation, or fibrous contraction
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Abnormal Pupil...Shape

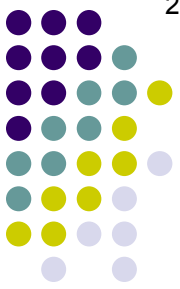
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Pupil structure abnormality

Abnormal Pupil...Location

Abnormal Pupil...Number

- Polycoria** = >1 pupil



Abnormalities of Pupil Structure

Abnormal Pupil...Size

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 - Due to dilator absence/malformation, or fibrous contraction
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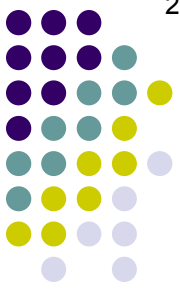
- Dyscoria** = congenital misshapen pupil
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Pupil structure abnormality

Abnormal Pupil...Location

Abnormal Pupil...Number

- Polycoria** = >1 pupil
- True polycoria** requires that each pupil has a...



Abnormalities of Pupil Structure

Abnormal Pupil...Size

- 1) Congenital **mios**is (*microcoria*)
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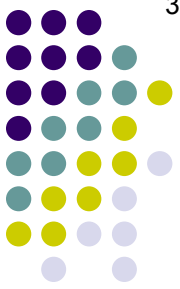
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Pupil structure abnormality

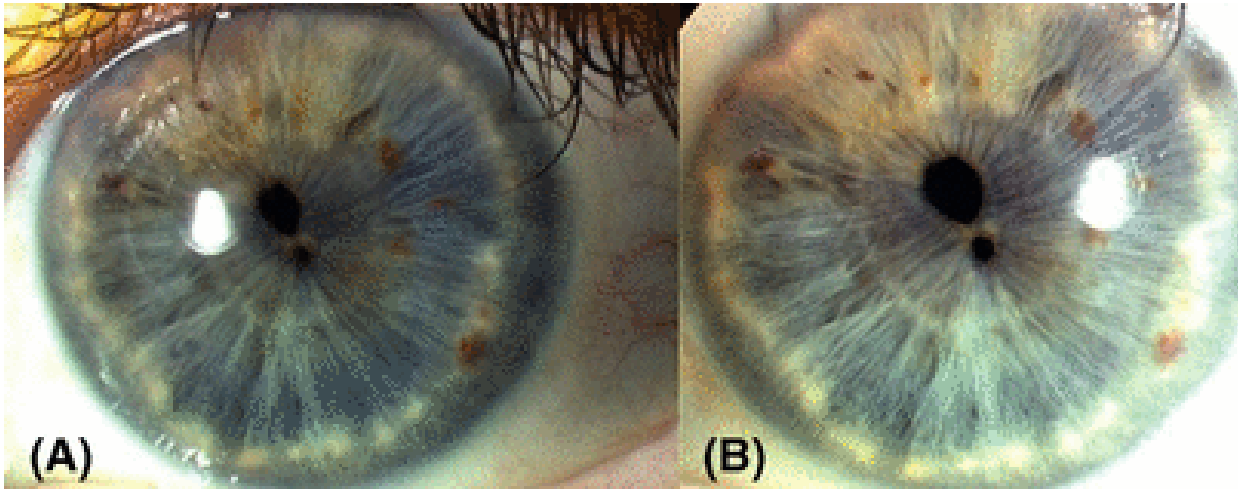
Abnormal Pupil...Location

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- Polycoria** = >1 pupil
- True polycoria** requires that each pupil has a...**sphincter** muscle (is exceedingly rare)

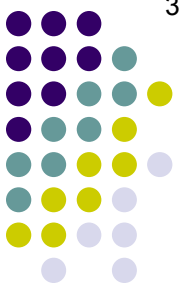


Abnormalities of Pupil Structure

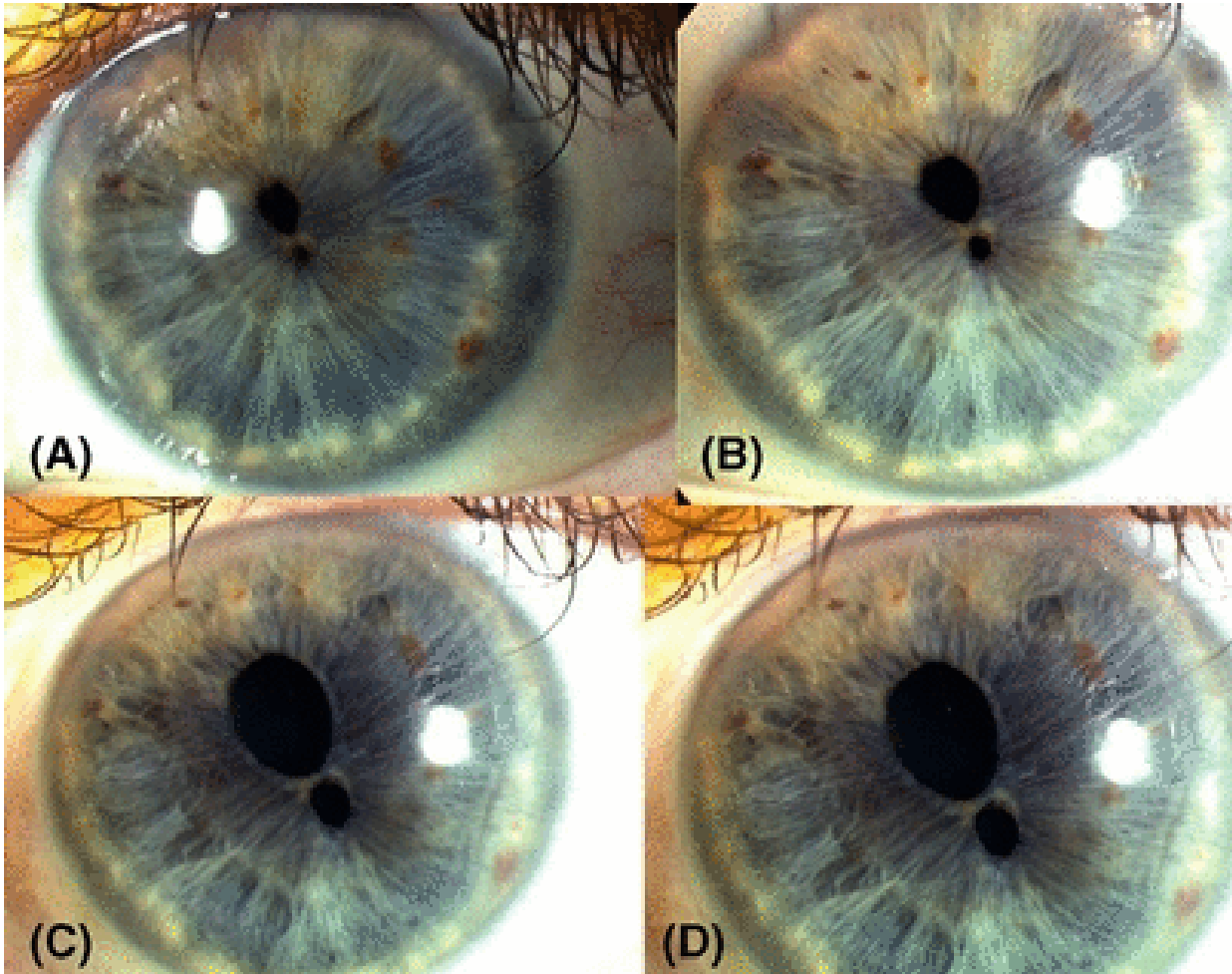


Eye in bright light (A) and dim light (B).

True polycoria

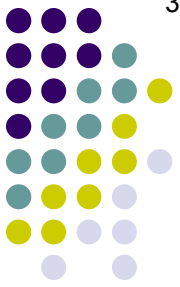


Abnormalities of Pupil Structure



Eye in bright light (A) and dim light (B).
Following dilatation with tropicamide and phenylephrine (C,D)

True polycoria



Abnormalities of Pupil Structure

Abnormal Pupil...Size

- 1) Congenital **mios**is (*microcoria*)
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Abnormal Pupil...Shape

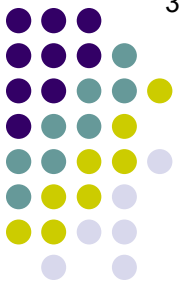
- Dyscoria** = congenital misshapen pupil
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Pupil structure abnormality

Abnormal Pupil...Location

Abnormal Pupil...Number

- Polycoria** = >1 pupil
- True polycoria** requires that each pupil has a...**sphincter muscle** (is exceedingly rare)
- accounts for the vast majority of cases



Abnormalities of Pupil Structure

Abnormal Pupil...Size

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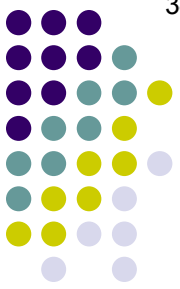
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Pupil structure abnormality

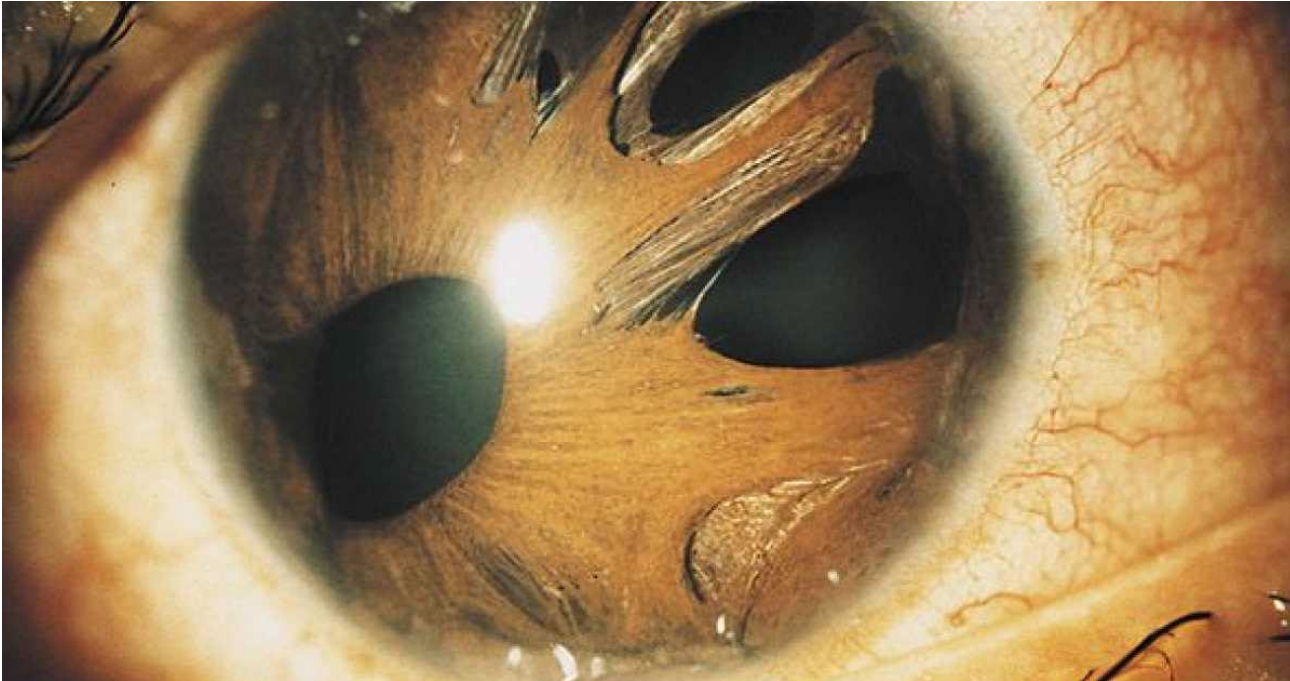
Abnormal Pupil...Location

Abnormal Pupil...Number

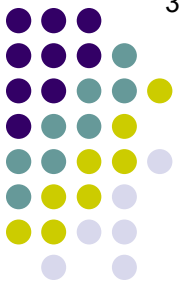
- Polycoria** = >1 pupil
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- Pseudopolycoria** accounts for the vast majority of cases



Abnormalities of Pupil Structure



Pseudopolyopia in ICE syndrome



Abnormalities of Pupil Structure

Abnormal Pupil...Size

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 - Associated with multiple congenital ocular abnormalities
- 2) Congenital **mydri**asis
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Abnormal Pupil...Shape

- Dyscoria** = congenital misshapen pupil
- e.g., coloboma

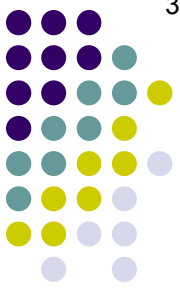
Pupil structure abnormality

Abnormal Pupil...Location

Corectopia =

Abnormal Pupil...Number

- Polycoria** = >1 pupil
- True polycoria** requires that each pupil has a...**sphincter** muscle (is exceedingly rare)
- Pseudopolycoria** accounts for the vast majority of cases



Abnormalities of Pupil Structure

Abnormal Pupil...Size

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Abnormal Pupil...Shape

- Dyscoria** = congenital misshapen pupil
- e.g., coloboma

Pupil structure abnormality

Abnormal Pupil...Location

Corectopia = displacement of pupil

Abnormal Pupil...Number

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- True polycoria** requires that each pupil has a...**sphincter muscle** (is exceedingly rare)
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Abnormalities of Pupil Structure

Abnormal Pupil...Size

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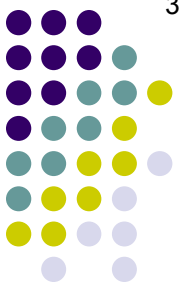
Pupil structure abnormality

Abnormal Pupil...Location

- Corectopia** = displacement of pupil
- Slight (.5mm) direction deviation is **normal**

Abnormal Pupil...Number

- Polycoria** = >1 pupil
- True polycoria** requires that each pupil has a...**sphincter muscle** (is exceedingly rare)
- Pseudopolycoria** accounts for the vast majority of cases



Abnormalities of Pupil Structure

Abnormal Pupil...Size

- 1) Congenital **miosis** (*microcoria*)
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 - Probably part of the aniridia spectrum

Abnormal Pupil...Shape

- Dyscoria** = congenital misshapen pupil
- e.g., coloboma

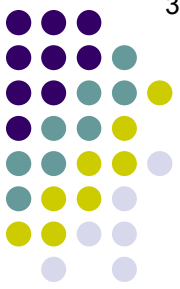
Pupil structure abnormality

Abnormal Pupil...Location

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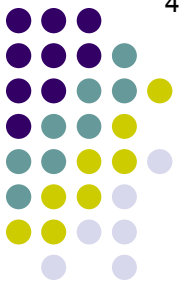
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Abnormalities of Pupil Structure



Corectopia



Abnormalities of Pupil Structure

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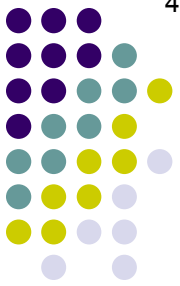
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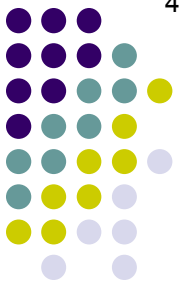
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Pupil structure abnormality

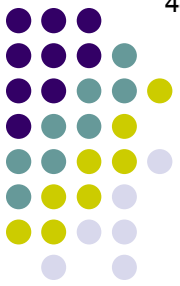
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- a) }
 - b) }
 - c) }
- 3 high-yield facts about *ectopia lentis et pupillae*

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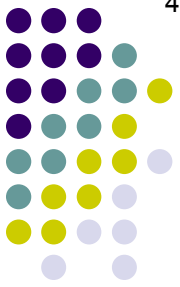
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- a) uni- v bilateral condition
 - b)
 - c)

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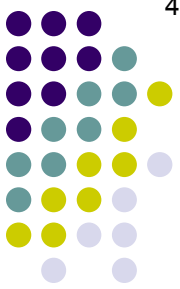
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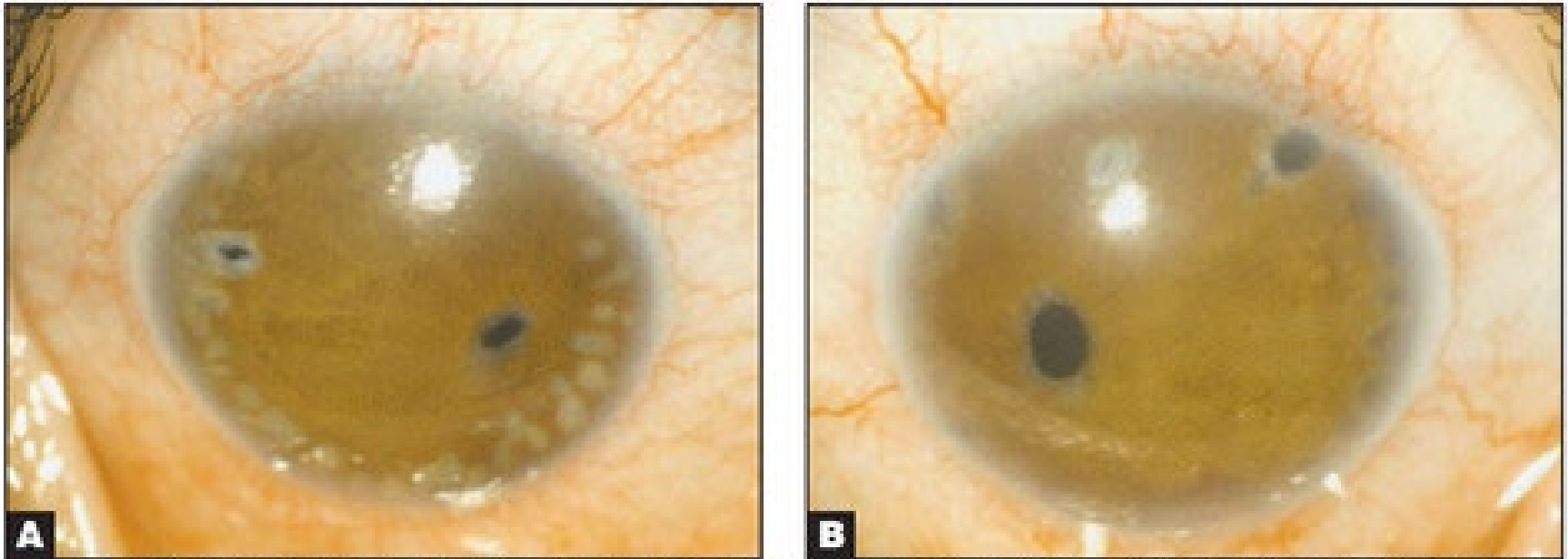
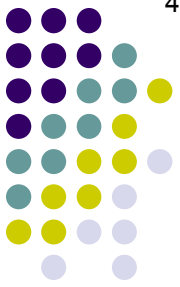


Figure 1. Preoperative view of both eyes in a patient with bilateral ectopia lentis et pupillae syndrome showing several lesions on the iris due to previous laser therapy. (A) Right eye. (B) Left eye.

Ectopia lentis et pupillae: Bilateral involvement



Abnormalities of Pupil Structure

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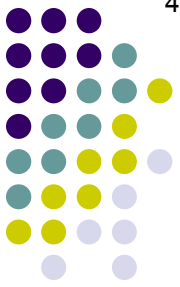
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 - a) Bilateral condition
 - b) Pupil and lens displaced in same vs opposite directions
 - c)

same vs
opposite

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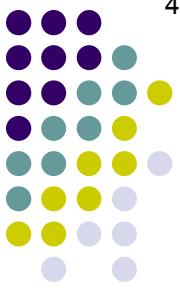
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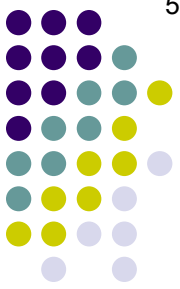
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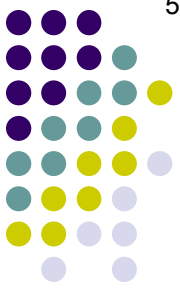
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Pupils inferotemporal, lenses superonasal

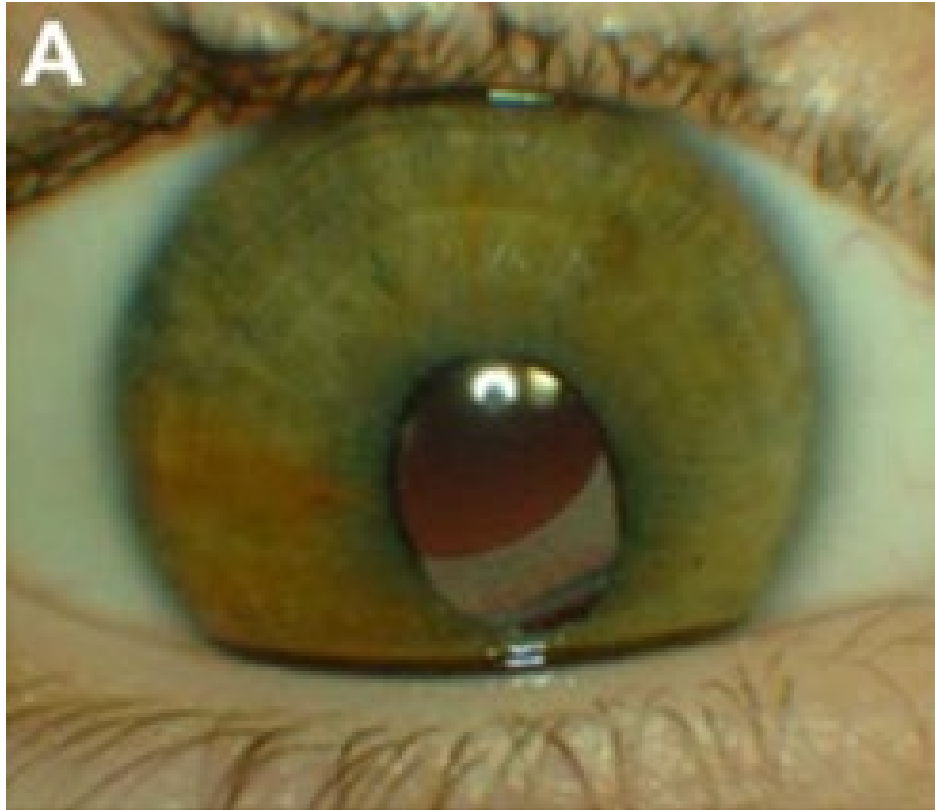
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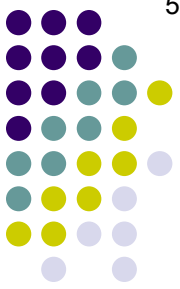
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Abnormalities of Pupil Structure



Ectopia lentis et pupillae: Pupil displaced inferonasal; lens, superotemporal



Abnormalities of Pupil Structure

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Pupil structure abnormality

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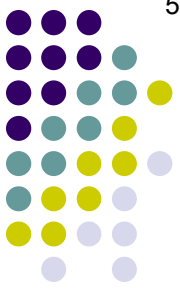
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Can the pupils and lenses be so displaced that the lens isn't located within the pupillary aperture?



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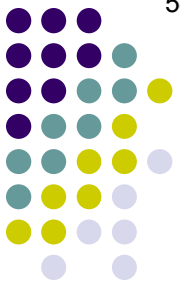
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They can indeed



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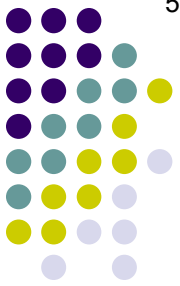
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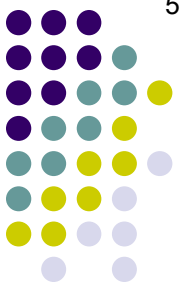
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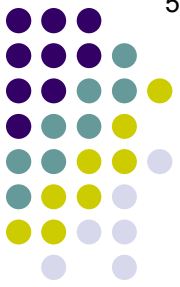
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Abnormalities of Pupil Structure



Ectopia lentis et pupillae: Bilateral inferonasal displacement of slit(ish) pupils



Abnormalities of Pupil Structure

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Pupil structure abnormality

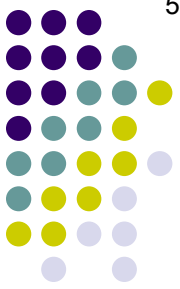
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Could have been listed up there as well



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Pupil structure abnormality

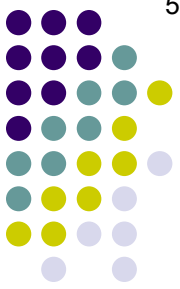
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Speaking of misshapen...What is the typical shape of the lens in ectopia lentis et pupillae?



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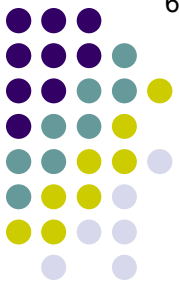
a) Bilateral condition

b) *Speaking of misshapen...What is the typical shape of the lens in ectopia lentis et pupillae?*

c) Microspherophakic

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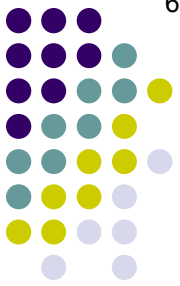
In a few words, how would you describe the shape of a microspherophakic lens?

Abnormal Pupil...Number

polyopia
polyopia requires that
...sphincter
(extremely rare)
polyopia accounts for
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Speaking of misshapen...what is the typical
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Microspherophakic



Abnormalities of Pupil Structure

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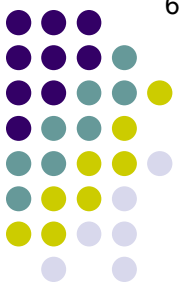
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The name says it all: the lens is small ('micro') and round ('sphero')

Abnormal Pupil...Number

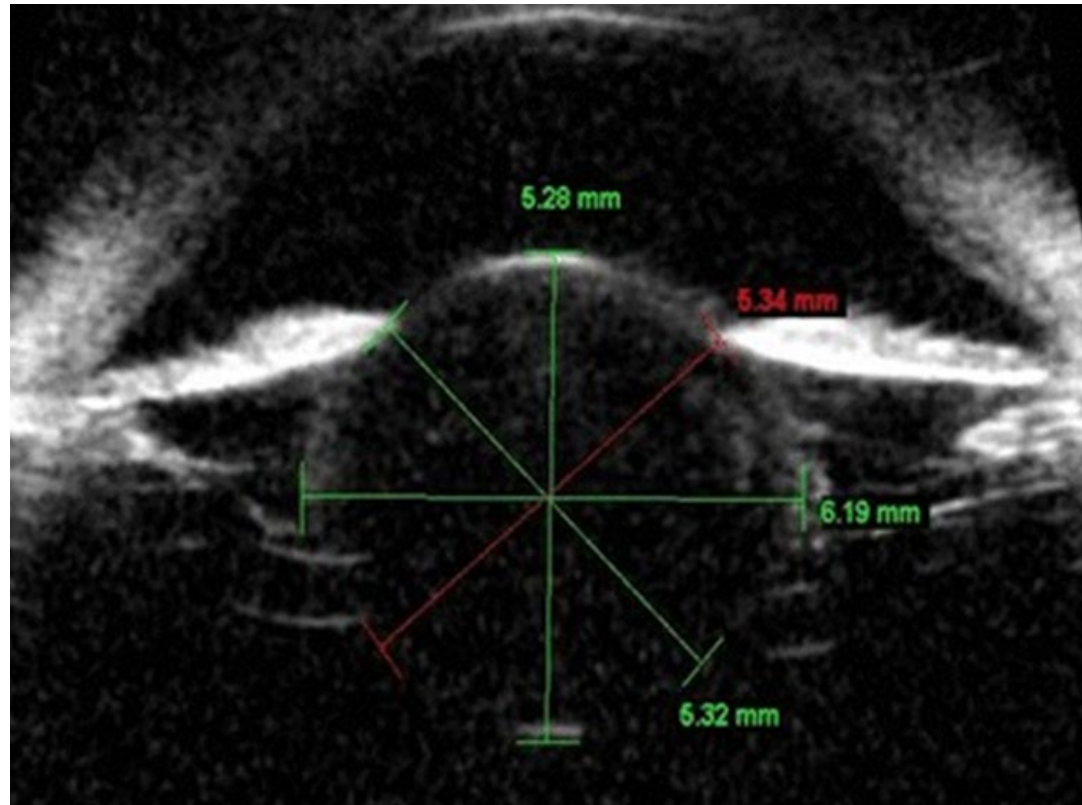
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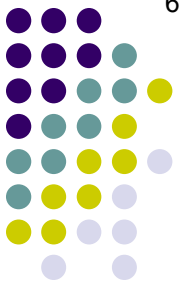
Microspherophakic



Abnormalities of Pupil Structure



Microspherophakia. Note the small size, round shape of the lens



Abnormalities of Pupil Structure

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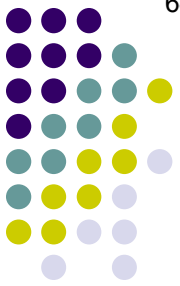
What common slit-lamp observation owes to the lens' small size?

Abnormal Pupil...Number

polyopia
pupil
requires that
...sphincter
(extremely rare)
polyopia accounts for
of cases

Speaking of misshapen...what is the typical shape of the lens in ectopia lentis et pupillae?

Microspherophakic



Abnormalities of Pupil Structure

Abnormal Pupil...Size

- 1) Congenital **miosis** (*microcoria*)
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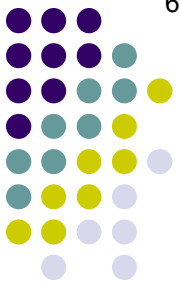
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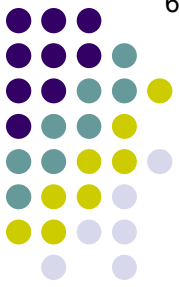
Microspherophakic



Abnormalities of Pupil Structure



Microspherophakia. Lens is able to fit through the pupillary aperture with mydriasis



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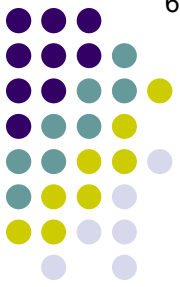
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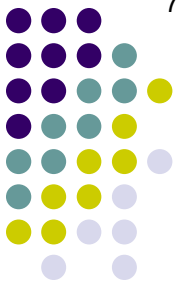
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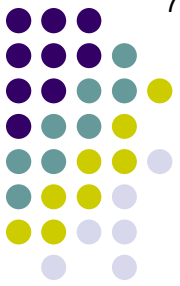
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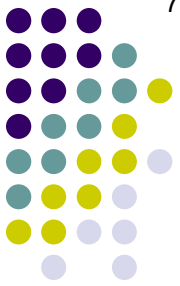
Weill-Marchesani syndrome

Abnormal Pupil...Number

Polypupils requires that the iris sphincter is abnormally (and rarely) duplicated (accounts for 1% of cases)

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Microspherophakic



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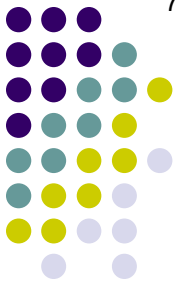
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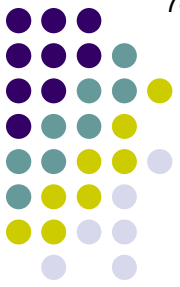
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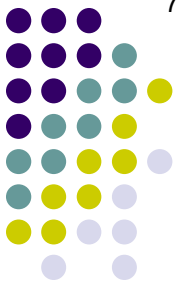
Microspherophakic



Abnormalities of Pupil Structure



Weill-Marchesani syndrome: Short stature



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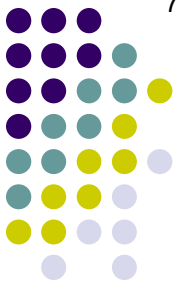
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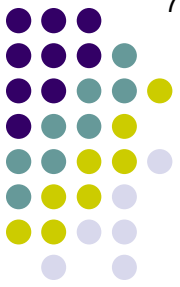
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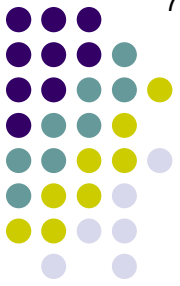
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Abnormalities of Pupil Structure



Weill-Marchesani syndrome: Short digits



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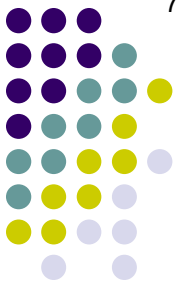
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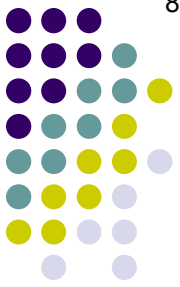
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For more on Weill-Marchesani, see slide-set FELT14

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