

## Q *Chronic Progressive External Ophthalmoplegia (CPEO)*



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# A *Chronic Progressive External Ophthalmoplegia (CPEO)*



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# A Chronic Progressive External Ophthalmoplegia (CPEO)



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## Q Chronic Progressive External Ophthalmoplegia (CPEO)



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  - It begins with ptosis, usually *in childhood* ~~after age 60~~ ~~F~~ T
  - It is a mitochondrial disease



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  - It is a mitochondrial disease **T**
  - Transmission can be maternal, AR, AD, or sporadic



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- Which of the following concerning CPEO are true?
  - It begins with ptosis, usually <sup>in childhood</sup> ~~after age 60~~ ~~F~~ T
  - It is a mitochondrial disease T
  - Transmission can be **maternal**, AR, AD, or sporadic T

*How does maternal transmission manifest?*



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*How does maternal transmission manifest?*

Only mothers can transmit the dz, and both male and female offspring can get it



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  - **It is a mitochondrial disease T**
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Huh? I thought mitochondrial diseases were always transmitted in maternal fashion.  
How can a mitochondrial (Mt) dz be transmitted AR or AD?



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*Huh? I thought mitochondrial diseases were always transmitted in maternal fashion. How can a mitochondrial (Mt) dz be transmitted AR or AD?*

Some of the proteins that influence Mt gene expression are coded for by nuclear DNA; ie, DNA of the cell in which the Mt resides. Thus, like other diseases that can result from coding errors in the nuclear DNA, CPEO can present in AR or AD fashion.



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*OK, but which form of transmission is most common?*

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*OK, but which form of transmission is most common?*  
None of the four BCSC books in which CPEO is discussed are definitive on this score. The *Neuro* book asserts that 'mtDNA point deletions' account for more cases than does nuclear DNA errors. However, it does not state whether those mtDNA deletions are inherited, or sporadic.



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  - Diplopia is a common complaint





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*Why is diplopia uncommon in CPEO?*



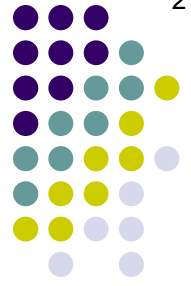
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  - It is associated with VF and ERG abnormalities T
  - Diplopia is <sup>an uncommon</sup> ~~a common~~ complaint ~~F~~ T

*Why is diplopia uncommon in CPEO?*

Because the ophthalmoplegia is usually symmetric bilaterally

## Chronic Progressive External Ophthalmoplegia (CPEO)



CPEO: Symmetric ophthalmoplegia



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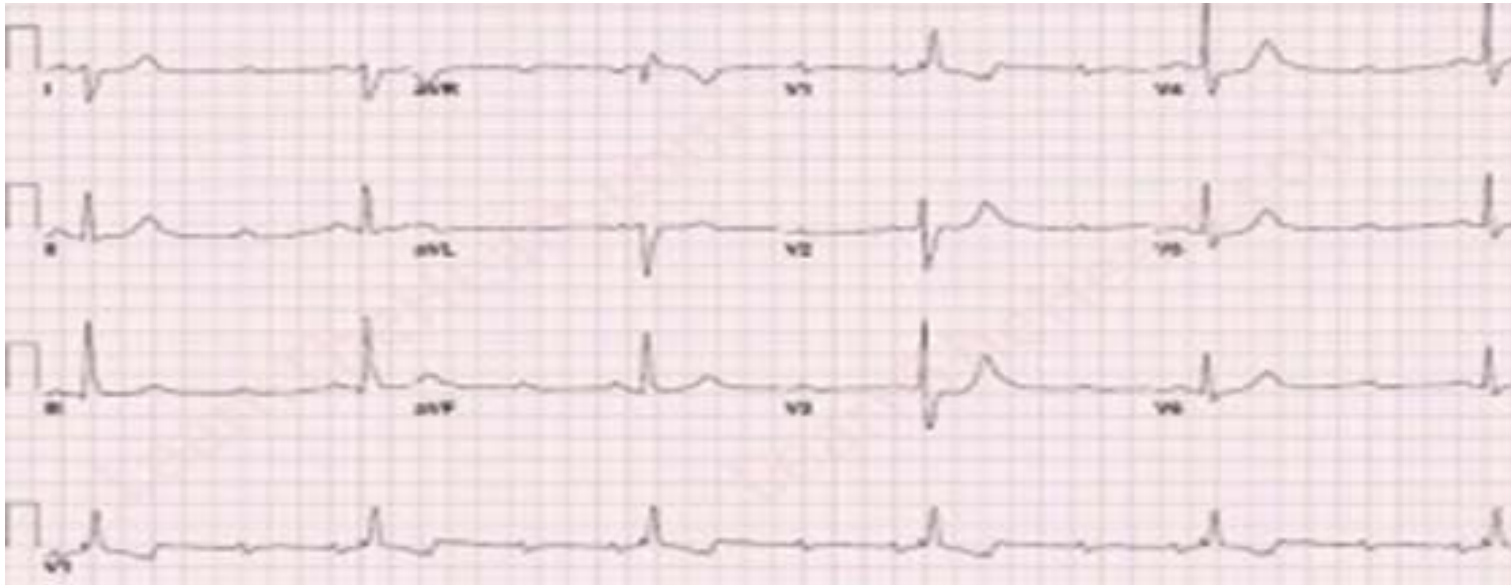
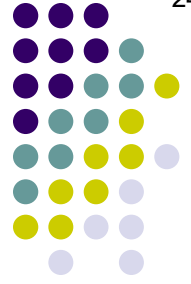
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  - Diplopia is *an uncommon* ~~a common~~ complaint ~~F~~ T
  - It may be associated with cardiac abnormalities



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## Chronic Progressive External Ophthalmoplegia (CPEO)



CPEO: Complete heart block in a 15 y.o.





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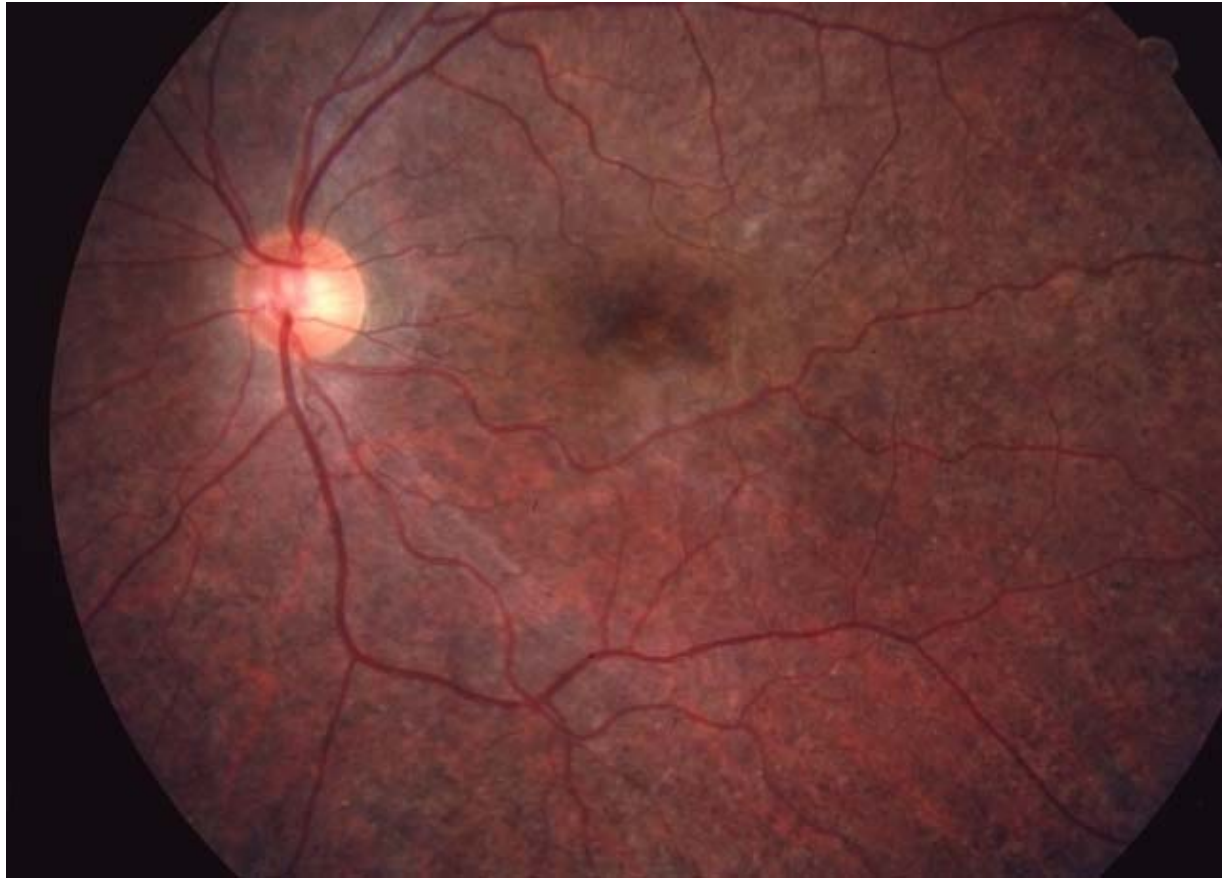
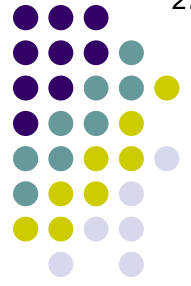
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  - It may be associated with cardiac abnormalities T
  - It may be associated with a pigmentary retinopathy



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## Chronic Progressive External Ophthalmoplegia (CPEO)



CPEO: Pigmentary retinopathy

## Q Chronic Progressive External Ophthalmoplegia (CPEO)

CPEO + pigmentary retinopathy + cardiac conduction problems = what syndrome?

- Diplopia is a common complaint
- It may be associated with **cardiac abnormalities**
- It may be associated with a **pigmentary retinopathy**

**A**

## Chronic Progressive External Ophthalmoplegia (CPEO)

CPEO + pigmentary retinopathy + cardiac conduction problems = what syndrome?  
Kearns-Sayre syndrome (KSS)

- Diplopia is a common complaint
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*CPEO + pigmentary retinopathy + cardiac conduction problems = what syndrome?  
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*Are the cardiac issues a source of significant morbidity?*

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Yes—they are often fatal (and are a large reason why KSS is associated with a markedly shortened lifespan)

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*Are the cardiac issues a source of significant morbidity?*

Yes—they are often fatal (and are a large reason why KSS is associated with a markedly shortened lifespan)

*Does ptosis/ophthalmoplegia precede, or follow the onset of cardiac issues?*

- Diplopia is ~~a common complaint~~
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Precede

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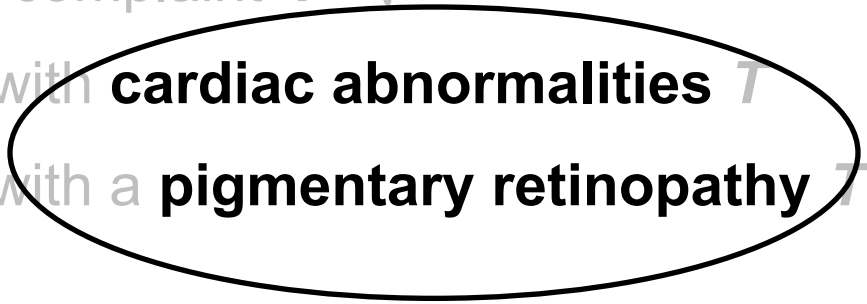
Yes—**they are often fatal** (and are a large reason why KSS is associated with a markedly shortened life expectancy)

What does this imply re management?

Does ptosis/corneal clouding precede cardiac issues?

## Precede

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*What does this imply re management?*

It implies that all cases of progressive ptosis+ophthalmoplegia should undergo **cardiac evaluation**

Does ptosis/ophthalmoplegia precede cardiac issues?

**Precede**

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- Diplopia is a common complaint T
- It may be associated with cardiac abnormalities T
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**A final note about KSS:** The latest copy (in my possession) of the *Fundamentals* book states KSS consists of CPEO, heart block and “severe RP.” The three other *BCSC* books that address this topic do not include ‘severe RP’ in their descriptions of KSS (although KSS is, *on account of its appearance*, in the DDX for an RP-like fundus). I think the *Fundamentals* book is in error on this score, but caveat emptor.



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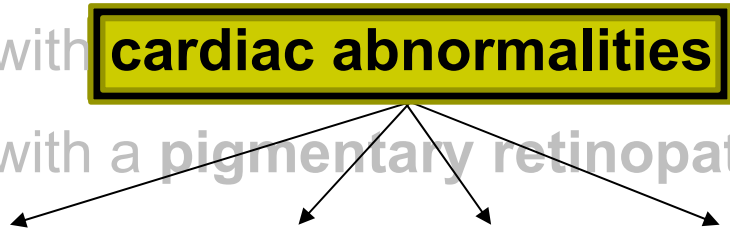
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Speaking of cardiac conduction issues—when an eye dentist encounters those words, four conditions should come to mind (although admittedly, one of them probably needn't stay there for long).

No question yet—proceed when ready



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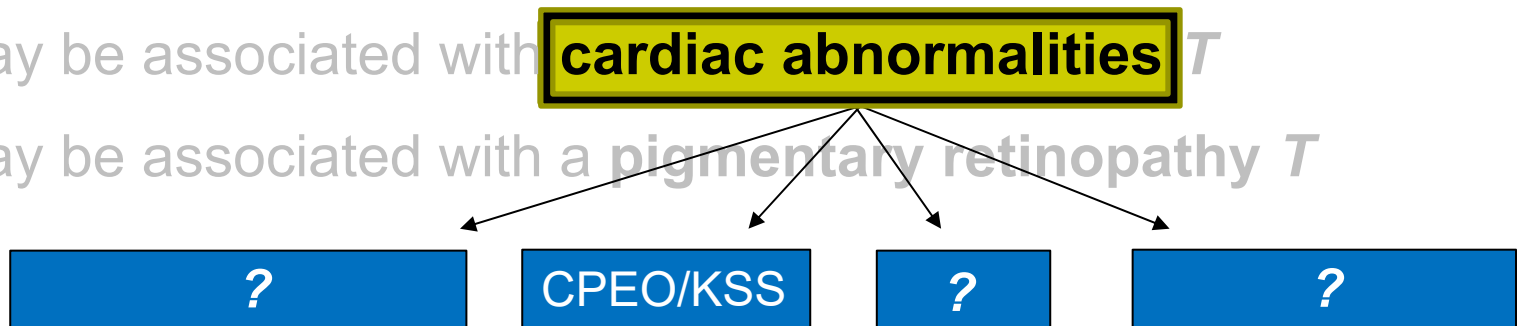
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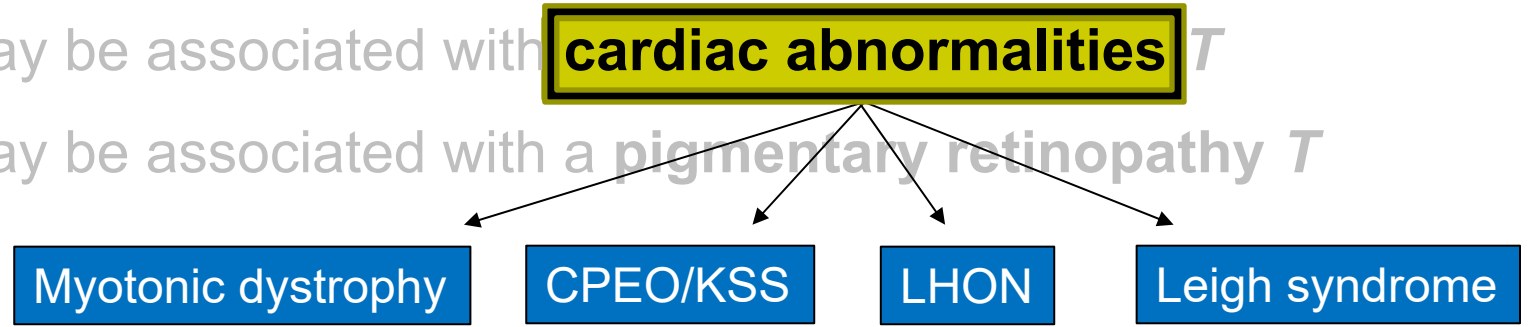
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In a nutshell, what sort of condition is myotonic dystrophy?

associated with a

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**cardiac abnormalities**

**Myotonic dystrophy**

**CPEO/KSS**

**LHON**

**Leigh syndrome**

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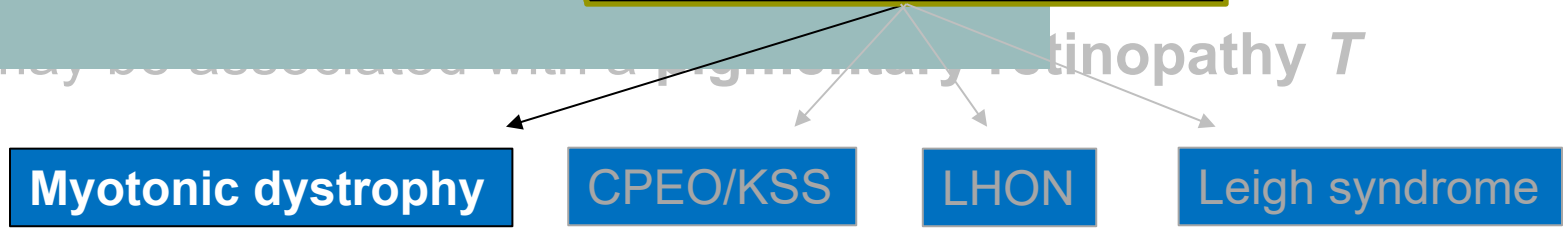
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What are its other ocular manifestations?

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- 
- 

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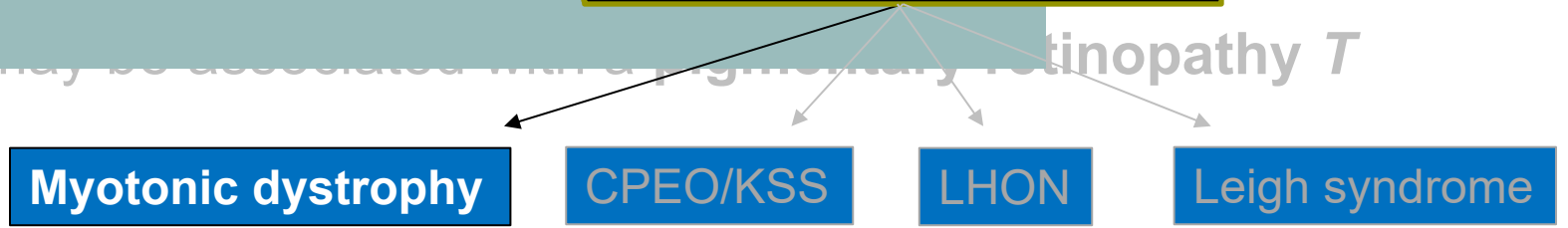
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- Pigmentary retinopathy
- two-word description cataracts

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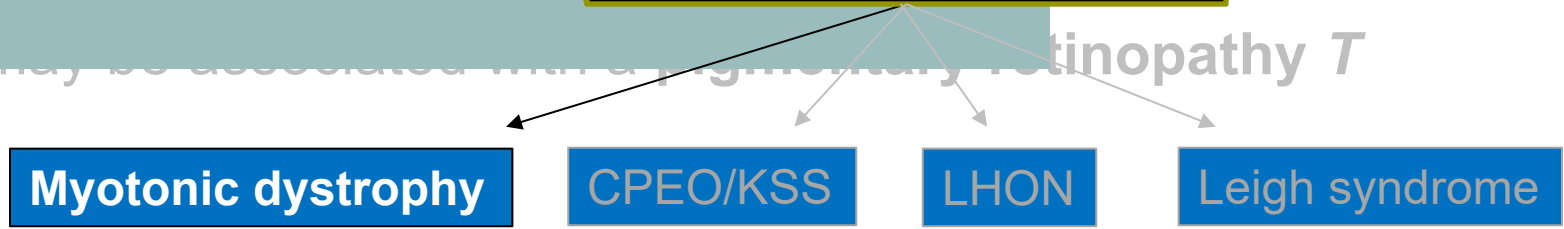
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*What are its classic nonocular findings?*

- Cardiac conduction issues
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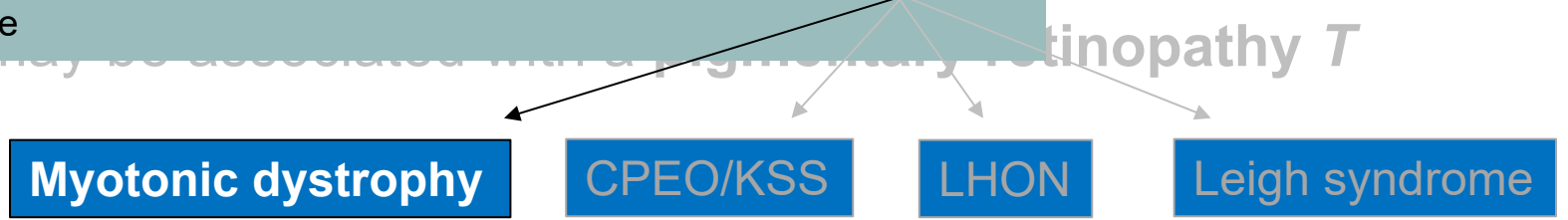
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- Cardiac conduction issues
- Myotonia
- Characteristic        facies
- Frontal balding
- Low intelligence

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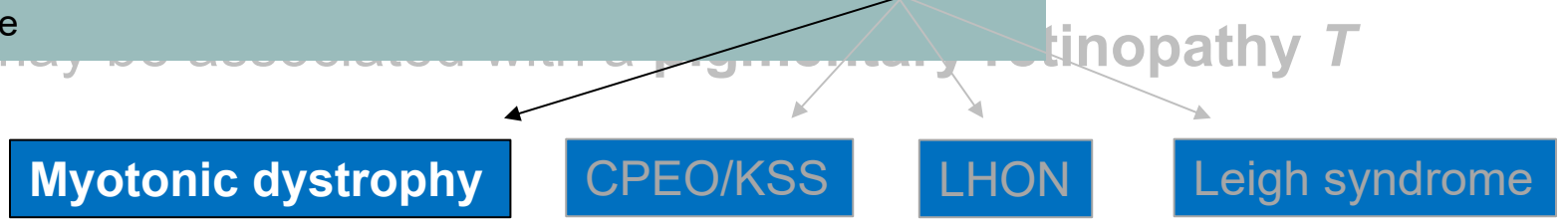
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- Characteristic 'hatchet' facies
- Frontal balding
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## cardiac abnormalities



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Are the cardiac issues a source of significant morbidity?

Yes—they are often fatal (and are a large reason why KSS is associated with a markedly shortened life expectancy)

What does LHON stand for in this context?

Does  
Pre

issues?

**cardiac abnormalities**

- It may be associated with a pigmentary retinopathy



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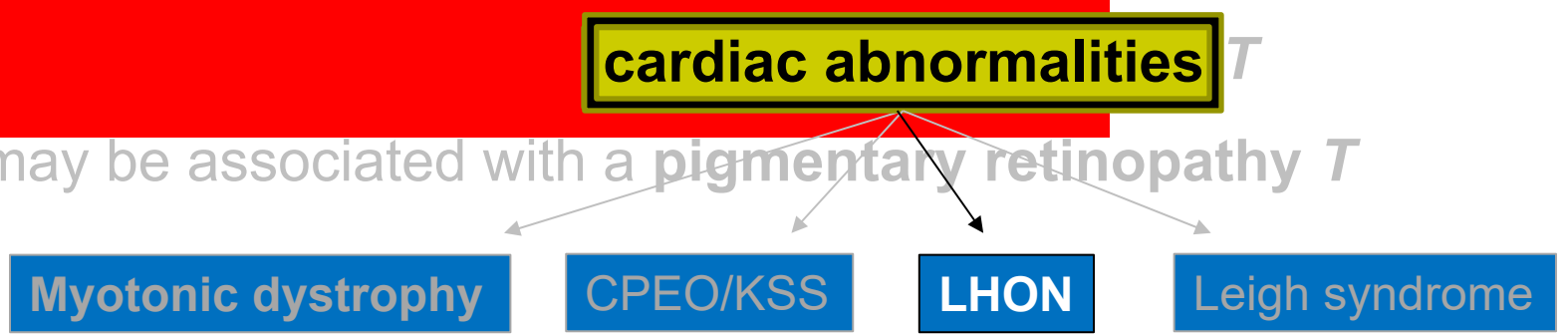
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What does LHON stand for in this context?  
Leber's hereditary optic neuropathy

Does it cause cardiac issues?  
Prevalence of cardiac issues?

- It may be associated with a pigmentary retinopathy



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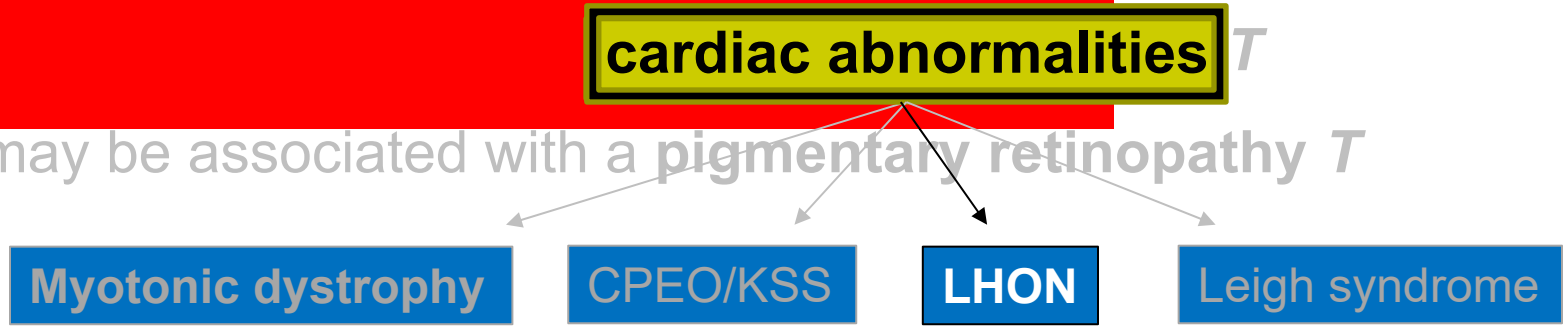
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Yes—they are often fatal (and are a large reason why KSS is associated with a markedly reduced life expectancy)

What does LHON stand for in this context?  
Leber's hereditary optic neuropathy

How does it present?

It may be associated with a pigmentary retinopathy



Speaking of cardiac conduction issues—when an eye dentist encounters those words, four conditions should come to mind (although admittedly, one of them probably needn't stay there for long). One is CPEO/KSS. What are the other three?



# A Chronic Progressive External Ophthalmoplegia (CPEO)

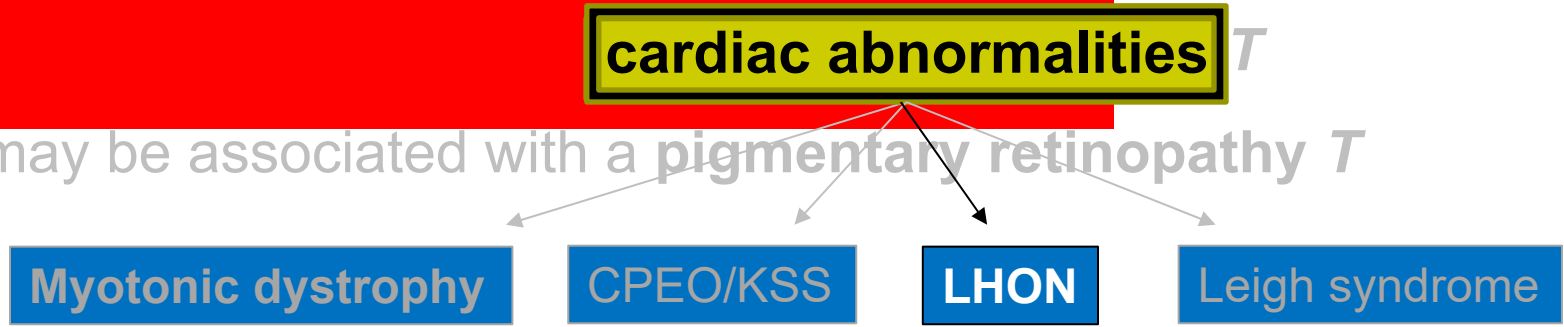
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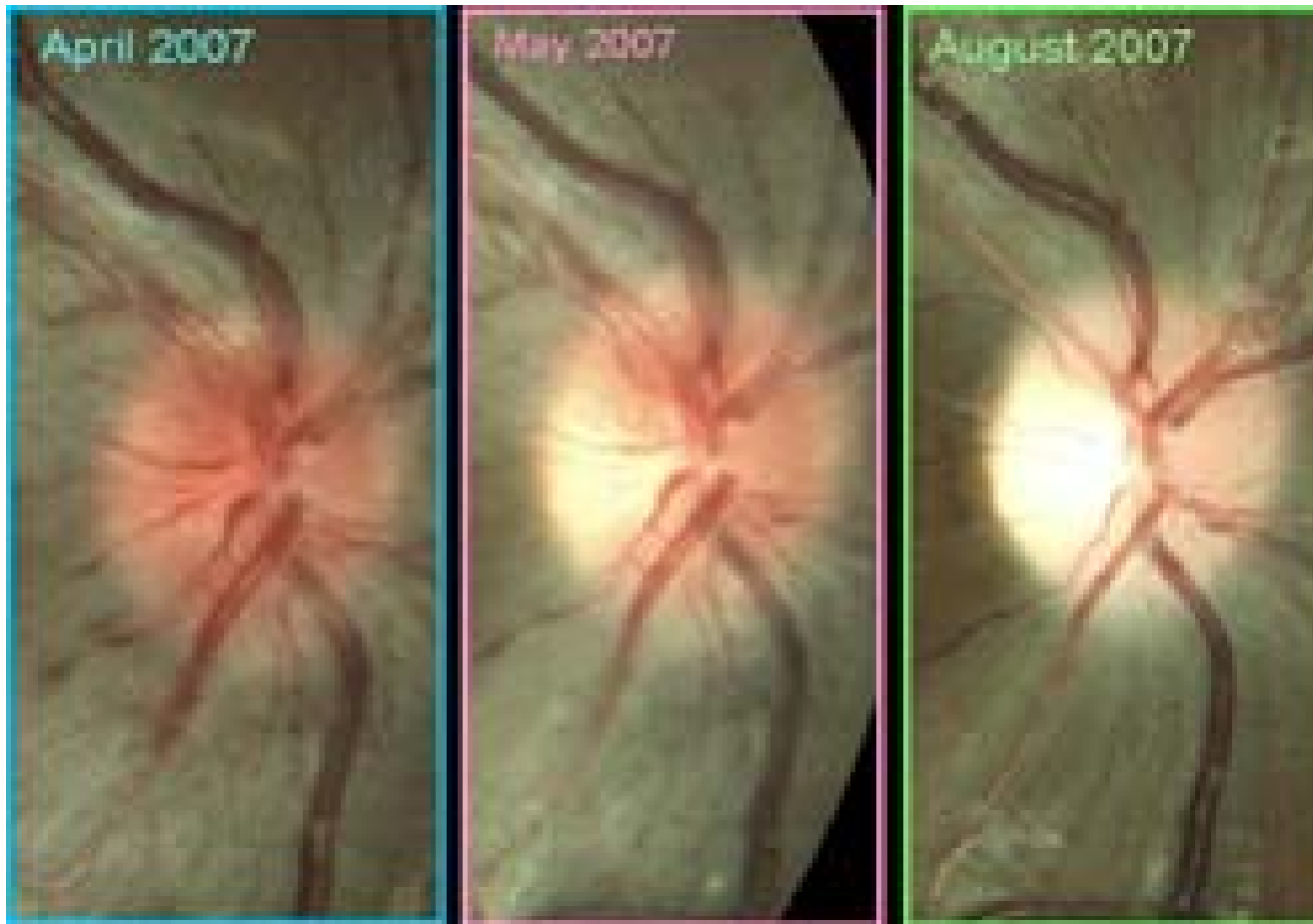
*How does it present?*  
With decreased vision (initially unilateral, but the fellow eye is inevitably affected as well) and optic atrophy in the second to fourth decade

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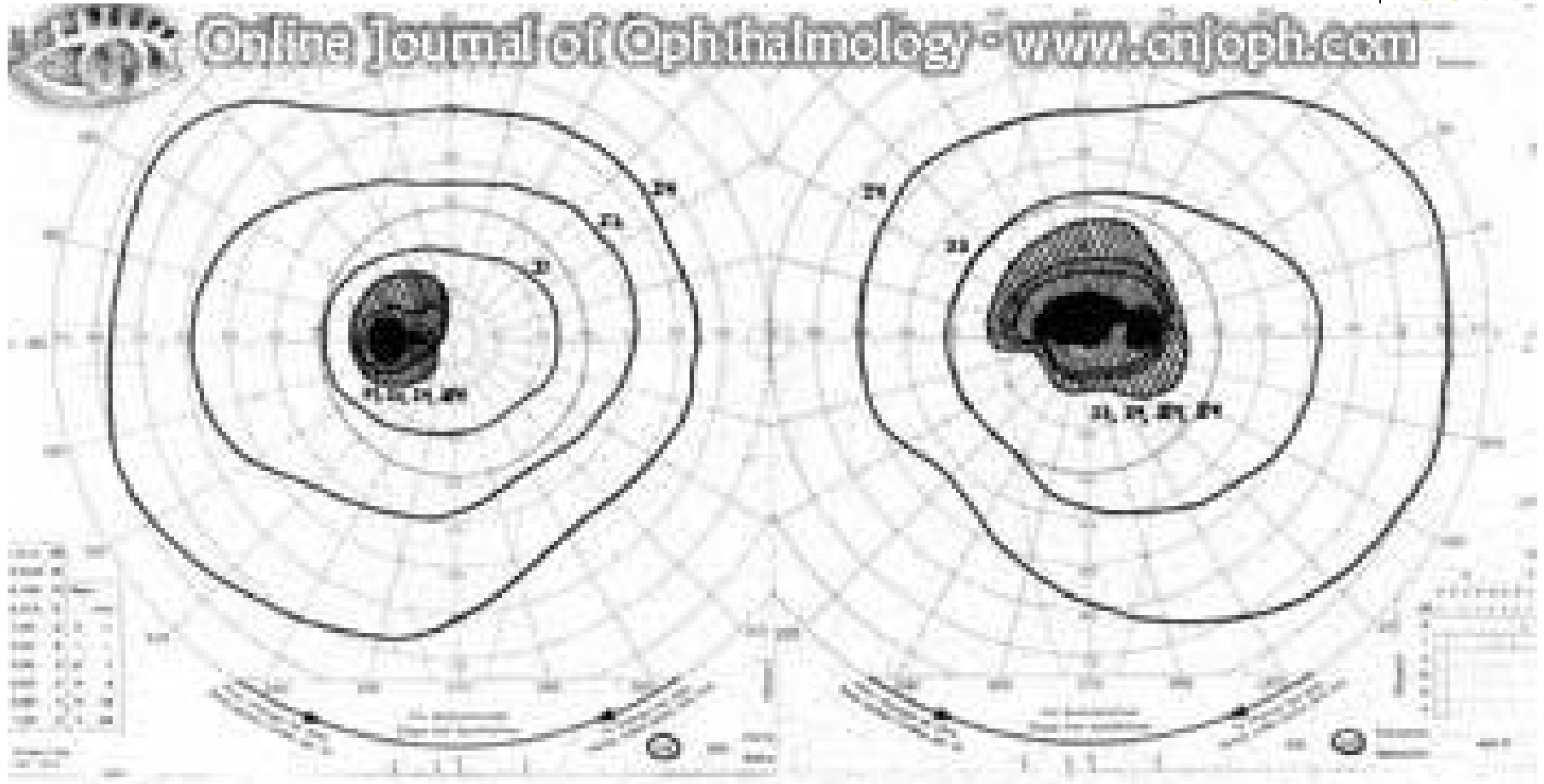
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## Chronic Progressive External Ophthalmoplegia (CPEO)



LHON: Progression of ONH atrophy

*Chronic Progressive External Ophthalmoplegia (CPEO)*



LHON: Central/cecocentral scotomata



# Q Chronic Progressive External Ophthalmoplegia (CPEO)

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Does it have a gender predilection?

**cardiac abnormalities**

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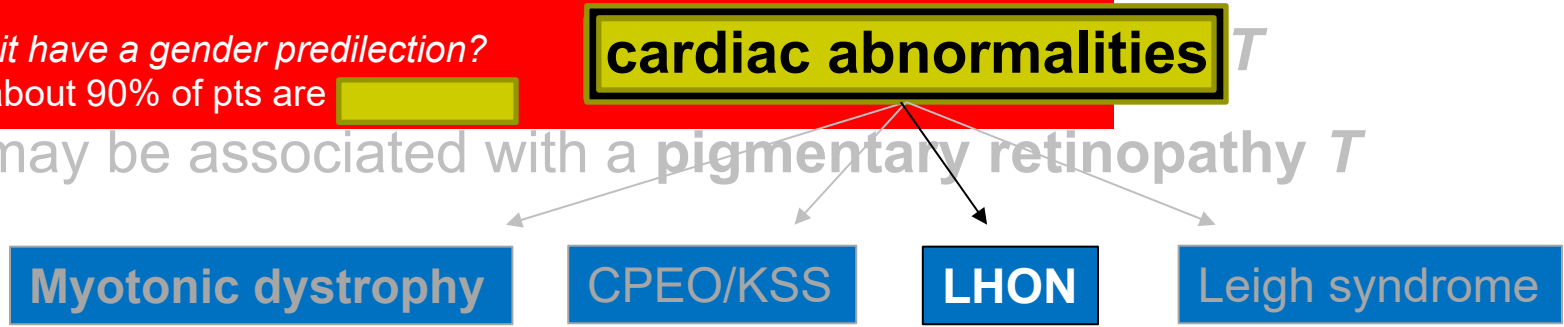
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**Does it have a gender predilection?**  
Yes, about 90% of pts are male

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What does this imply re management?  
It implies that all cases of progressive ptosis+ophthalmoplegia should undergo cardiac evaluation

Does ptosis/ophthalmoplegia precede cardiac issues?  
Precede

- Diplopia is a common complaint
- It may be associated with cardiac abnormalities
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Leigh syndrome is the one you can probably forget. (It has a full entry in Eyewiki, but receives only one mention—in a Table—in the BCSC.)

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Leigh syndrome

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## Q Chronic Progressive External Ophthalmoplegia (CPEO)

- Which of the following concerning CPEO are true?
  - It begins with ptosis, usually *in childhood* ~~after age 60~~ ~~F~~ **T**
  - It is a mitochondrial disease **T**
  - Transmission can be maternal, AR, AD, or sporadic **T**
  - It is associated with VF and ERG abnormalities **T**
  - Diplopia is *an uncommon* ~~a common~~ complaint ~~F~~ **T**
  - It may be associated with cardiac abnormalities **T**
  - It may be associated with a pigmentary retinopathy **T**
  - The pupil is involved



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*Why is this a really dumb question? That is, why is it painfully obvious that CPEO is, by definition, pupil sparing?*

- It may be associated with a pigmentary retinopathy /

• The pupil is <sup>not</sup> involved ~~F~~ T

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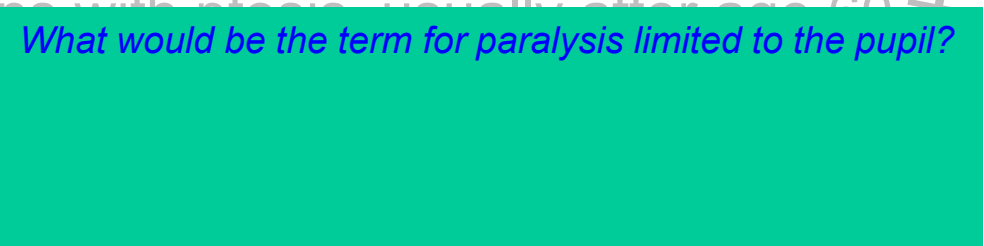
The answer is right there in the name. The term *external ophthalmoplegia* refers to paralysis of *extraocular* muscles--the prefix *extra* here meaning 'external to the globe itself.' Obviously, the internally-located pupil cannot be involved in such a process.

- It may be associated with a pigmentary retinopathy T

• The pupil is <sup>not</sup> involved ~~F~~ T



# Q Chronic Progressive External Ophthalmoplegia (CPEO)

- Which of the following concerning CPEO are true?
  - It begins with ptosis usually after age 50 *in childhood* ~~F~~ **T**
  - It is a 
  - Transmitted as an autosomal recessive trait *sporadic* ~~T~~
  - It is associated with VF and ERG abnormalities ~~T~~

*What would be the term for paralysis limited to the pupil?*

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- It may be associated with a pigmentary retinopathy ~~T~~

The pupil is <sup>not</sup> involved ~~F~~ **T**







# A Chronic Progressive External Ophthalmoplegia (CPEO)

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  - It begins with ptosis usually after age 50 *in childhood* ~~F~~ **T**
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The pupil is <sup>not</sup> involved ~~F~~ **T**





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- It is a **What would be the term for paralysis limited to the pupil?  
An internal ophthalmoplegia**

- Transient **What if the paralysis involves the EOMs and the pupil?**

- Transient or sporadic **T**

- It is associated with VF and ERG abnormalities **T**

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- It may be associated with a pigmentary retinopathy **T**

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# A Chronic Progressive External Ophthalmoplegia (CPEO)

- Which of the following concerning CPEO are true?
  - It begins *usually after age 50* *in childhood*
  - It is a *sporadic* *transient*
  - Trans *sporadic*
  - It is associated with VF and ERG abnormalities *T*

What would be the term for paralysis limited to the pupil?  
 An **internal** ophthalmoplegia

What if the paralysis involves the EOMs **and** the pupil?  
 A **complete** ophthalmoplegia

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What does muscle biopsy reveal?



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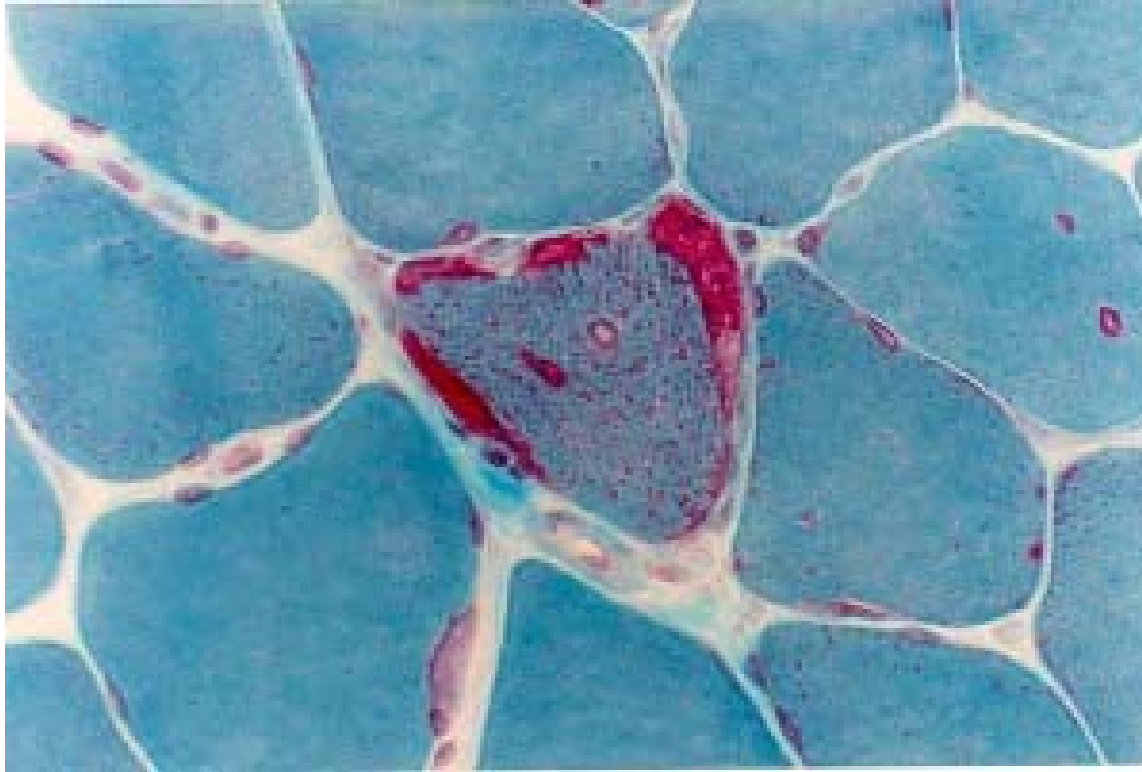
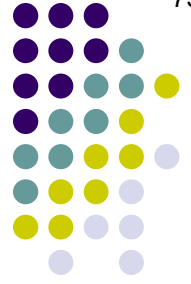
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*What does muscle biopsy reveal?*

The classic **ragged red fibers** you heard about in med school



## Chronic Progressive External Ophthalmoplegia (CPEO)



CPEO: 'Ragged red fibers'



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  - The *not* **Being able to differentiate CPEO from MG is important, so let's drill down on this**
  - It is **Myasthenia gravis (MG) is in the DDx for CPEO** T

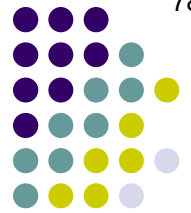


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	MG	CPEO
Fatigable?	?	?

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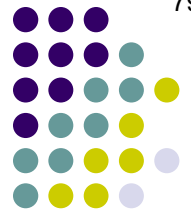


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What does it mean to say MG is fatigable?

- It may be associated with a pigmentary retinopathy T
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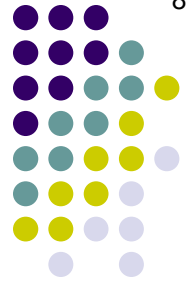
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What does it mean to say MG is fatigable?  
 It means (with regard to ptosis) that sustained lid elevation will lead to further weakening (ie, will *fatigue*) of the levator, causing the ptosis to worsen

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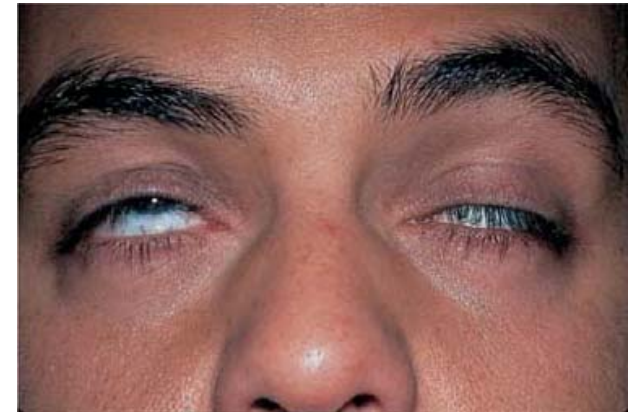
## Chronic Progressive External Ophthalmoplegia (CPEO)



In primary



After sustained upgaze



After further  
sustained upgaze

MG: Fatigability



**Q** *Chronic Progressive External Ophthalmoplegia (CPEO)*

- Which of the following concerning CPEO are true?
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	MG	CPEO
Fatigable?	Yes	No
+Tensilon test?	?	?

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What is the trade name for Tensilon?



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 Edrophonium (so this is aka the edrophonium test)

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Pharmacologically, what is edrophonium?

- Myasthenia gravis (MG) is in the DDx for CPEO **T**



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	MG	CPEO
<i>Fatigable?</i>	Yes	No
<b>+Tensilon test?</b>	<b>Yes</b>	No

*What is the trade name for Tensilon?  
Edrophonium (so this is aka the edrophonium test)*

*Pharmacologically, what is edrophonium?  
An acetylcholinesterase inhibitor*

- Myasthenia gravis (MG) is in the DDx for CPEO T**



## Q Chronic Progressive External Ophthalmoplegia (CPEO)

- Which of the following concerning CPEO are true?
  - It begins with ptosis, usually <sup>in childhood</sup> ~~after age 60~~ **F** **T**

	MG	CPEO
<i>Fatigable?</i>	Yes	No
<b>+Tensilon test?</b>	<b>Yes</b>	No

*What is the trade name for Tensilon?*

Edrophonium (so this is aka the *edrophonium test*)

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*Is it short-, or long-acting?*

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 Short (quite)

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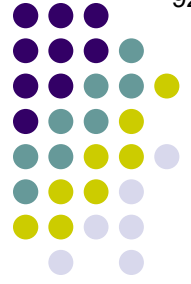
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*Is it short-, or long-acting?*  
 Short (quite)

*What constitutes a positive test?*  
 The temporary amelioration of MG S/S

- Myasthenia gravis (MG) is in the DDx for CPEO T**

## Chronic Progressive External Ophthalmoplegia (CPEO)



Pre-injection

A few seconds post-injection

MG: Tensilon test



# Q Chronic Progressive External Ophthalmoplegia (CPEO)

- Which of the following concerning CPEO are true?
  - It begins with ptosis, usually <sup>in childhood</sup> ~~after age 60~~ **F T**

	MG	CPEO
<i>Fatigable?</i>	Yes	No
<b>+Tensilon test?</b>	<b>Yes</b>	No

*Tensilon testing is not commonly performed, in part at least because of its potential adverse effects. Of these, which are most disconcerting?*

*Pharmacologically, what is edrophonium?*  
 An acetylcholinesterase inhibitor

*Is it short-, or long-acting?*  
 Short (quite)

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	MG	CPEO
<i>Fatigable?</i>	Yes	No
<b>+Tensilon test?</b>	<b>Yes</b>	No

Tensilon testing is not commonly performed, in part at least because of its potential adverse effects. Of these, which are most disconcerting?  
Cardiac and respiratory arrest

*Pharmacologically, what is edrophonium?*  
An acetylcholinesterase inhibitor

*Is it short-, or long-acting?*  
Short (quite)

*What constitutes a positive test?*  
The temporary amelioration of MG S/S

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  - It begins with ptosis, usually ~~after age 60~~ <sup>in childhood</sup> ~~F~~ T

	MG	CPEO
Fatigable?	Yes	No
+Tensilon test?	Yes	No
Clinical course	?	?

- Diplopia is ~~a common~~ <sup>an uncommon</sup> complaint ~~F~~ T
- It may be associated with cardiac abnormalities T
- It may be associated with a pigmentary retinopathy T
- The pupil is <sup>not</sup> involved ~~F~~ T
- It is diagnosed via ~~serology~~ <sup>muscle biopsy</sup> ~~F~~ T
- Myasthenia gravis (MG) is in the DDx for CPEO T**



# A Chronic Progressive External Ophthalmoplegia (CPEO)

- Which of the following concerning CPEO are true?
  - It begins with ptosis, usually ~~after age 60~~ <sup>in childhood</sup> ~~F~~ T

	MG	CPEO
Fatigable?	Yes	No
+Tensilon test?	Yes	No
Clinical course	Waxes and wanes	Progressive

- Diplopia is ~~a common~~ <sup>an uncommon</sup> complaint ~~F~~ T
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# Chronic Progressive External Ophthalmoplegia (CPEO)

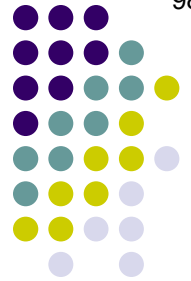
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  - It begins with ptosis, usually ~~after age 60~~ <sup>in childhood</sup> ~~F~~ T

	MG	CPEO
Fatigable?	Yes	No
+Tensilon test?	Yes	No
<b>Clinical course</b>	<b>Waxes and wanes</b>	<b>Progressive</b>

Of all the signs/symptoms, the *Neuro* book seems to emphasize this one in differentiating between MG and CPEO—the fact that MG waxes and wanes, whereas **CPEO is relentlessly progressive.**

- It may be associated with a pigmentary retinopathy T
- The pupil is <sup>not</sup> involved ~~F~~ T
- It is diagnosed via <sup>muscle biopsy</sup> ~~serology~~ ~~F~~ T
- Myasthenia gravis (MG) is in the DDx for CPEO T**

## Chronic Progressive External Ophthalmoplegia (CPEO)



1998



1999



2000



2002



2003



2004



Erin O'Malley, MD  
U of Iowa 2004

CPEO: Progressive ptosis

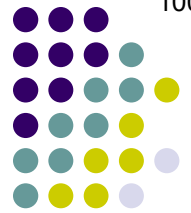


**Q** *Chronic Progressive External Ophthalmoplegia (CPEO)*

- Which of the following concerning CPEO are true?
  - It begins with ptosis, usually ~~after age 60~~ <sup>in childhood</sup> ~~F~~ **T**

	MG	CPEO
<i>Fatigable?</i>	Yes	No
<i>+Tensilon test?</i>	Yes	No
<i>Clinical course</i>	Waxes and wanes	Progressive
<i>Diplopia?</i>	?	?

- It may be associated with cardiac abnormalities **T**
- It may be associated with a pigmentary retinopathy **T**
- The pupil is <sup>not</sup> involved ~~F~~ **T**
- It is diagnosed via <sup>muscle biopsy</sup> ~~serology~~ ~~F~~ **T**
- Myasthenia gravis (MG) is in the DDx for CPEO **T****



# A Chronic Progressive External Ophthalmoplegia (CPEO)

- Which of the following concerning CPEO are true?
  - It begins with ptosis, usually ~~after age 60~~ <sup>in childhood</sup> ~~F~~ T

	MG	CPEO
Fatigable?	Yes	No
+Tensilon test?	Yes	No
Clinical course	Waxes and wanes	Progressive
Diplopia?	Yes	No

- It may be associated with cardiac abnormalities T
- It may be associated with a pigmentary retinopathy T
- The pupil is <sup>not</sup> involved ~~F~~ T
- It is diagnosed via <sup>muscle biopsy</sup> ~~serology~~ ~~F~~ T
- Myasthenia gravis (MG) is in the DDx for CPEO T**

## Chronic Progressive External Ophthalmoplegia (CPEO)

A note on this table: It is based on one in the *Peds/Strab* book. However, that table includes another condition to be considered, that being **thyroid eye disease (TED)**. For the life of me I cannot determine why TED is included—it causes eyelid **retraction**, not **ptosis**. If someone can explain the inclusion of TED to me, please do.

	MG	CPEO	TED
<i>Fatigable?</i>	Yes	No	No
<i>+Tensilon test?</i>	Yes	No	No
<i>Clinical course</i>	Waxes and wanes	Progressive	Variable—may progress, or resolve
<i>Diplopia?</i>	Yes	No	No

- It may be associated with cardiac abnormalities *T*
- It may be associated with a pigmentary retinopathy *T*
- The pupil is <sup>not</sup> involved ~~*F*~~ *T*
- It is diagnosed via <sup>muscle biopsy</sup> ~~serology~~ ~~*F*~~ *T*
- **Myasthenia gravis (MG) is in the DDx for CPEO** *T*

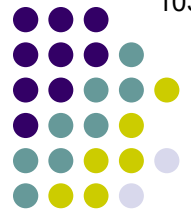


**Q** *Chronic Progressive External Ophthalmoplegia (CPEO)*

Which of the following concerning CPEO are true?  
 There is another condition—much less common than MG—that is frequently initially (mis)diagnosed as CPEO, and thus should be addressed here. What is it?

	MG	CPEO	?
Fatigable?	Yes	No	
+Tensilon test?	Yes	No	
Clinical course	Waxes and wanes	Progressive	
Diplopia?	Yes	No	

- It may be associated with cardiac abnormalities *T*
- It may be associated with a pigmentary retinopathy *T*
- The pupil is <sup>not</sup> involved ~~F~~ *T*
- It is diagnosed via <sup>muscle biopsy</sup> ~~serology~~ ~~F~~ *T*
- **Myasthenia gravis (MG) is in the DDx for CPEO** *T*



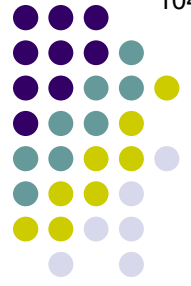
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Which of the following concerning CPEO are true?  
 There is another condition—much less common than MG—that is frequently initially (mis)diagnosed as CPEO, and thus should be addressed here. What is it?

	MG	CPEO	Myotonic Dystrophy
Fatigable?	Yes	No	
+Tensilon test?	Yes	No	
Clinical course	Waxes and wanes	Progressive	
Diplopia?	Yes	No	

- It may be associated with cardiac abnormalities *T*
- It may be associated with a pigmentary retinopathy *T*
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## Chronic Progressive External Ophthalmoplegia (CPEO)



Myotonic dystrophy



## Q Chronic Progressive External Ophthalmoplegia (CPEO)

In a nutshell, what sort of condition is myotonic dystrophy?

	MG	CPEO	Myotonic Dystrophy
Fatigable?	Yes	No	
+Tensilon test?	Yes	No	
Clinical course	Waxes and wanes	Progressive	
Diplopia?	Yes	No	

- It may be associated with cardiac abnormalities *T*
- It may be associated with a pigmentary retinopathy *T*
- The pupil is <sup>not</sup> involved ~~*F*~~ *T*
- It is diagnosed via <sup>muscle biopsy</sup> ~~serology~~ ~~*F*~~ *T*
- **Myasthenia gravis (MG) is in the DDX for CPEO** *T*

## Q/A Chronic Progressive External Ophthalmoplegia (CPEO)

In a nutshell, what sort of condition is myotonic dystrophy?

An inherited ( ) progressive systemic condition that results in ophthalmoplegia

AD?  
AR?  
XL?

	MG	CPEO	Myotonic Dystrophy
Fatigable?	Yes	No	
+Tensilon test?	Yes	No	
Clinical course	Waxes and wanes	Progressive	
Diplopia?	Yes	No	

- It may be associated with cardiac abnormalities *T*
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- **Myasthenia gravis (MG) is in the DDx for CPEO** *T*

# A Chronic Progressive External Ophthalmoplegia (CPEO)

In a nutshell, what sort of condition is myotonic dystrophy?

An inherited ( AD ) progressive systemic condition that results in ophthalmoplegia

	MG	CPEO	Myotonic Dystrophy
Fatigable?	Yes	No	
+Tensilon test?	Yes	No	
Clinical course	Waxes and wanes	Progressive	
Diplopia?	Yes	No	

- It may be associated with cardiac abnormalities *T*
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- The pupil is <sup>not</sup> involved ~~F~~ *T*
- It is diagnosed via <sup>muscle biopsy</sup> ~~serology~~ ~~F~~ *T*
- **Myasthenia gravis (MG) is in the DDx for CPEO** *T*



# A *Chronic Progressive External Ophthalmoplegia (CPEO)*

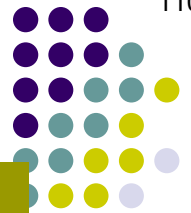
*In a nutshell, what sort of condition is myotonic dystrophy?*

An inherited ( AD ) progressive systemic condition that results in ophthalmoplegia

*Myotonic dystrophy has a number of manifestations—identify them.*

	<b>MG</b>	<b>CPEO</b>	<b>Myotonic Dystrophy</b>
<i>Fatigable?</i>	Yes	No	
<i>+Tensilon test?</i>	Yes	No	
<i>Clinical course</i>	Waxes and wanes	Progressive	
<i>Diplopia?</i>	Yes	No	
<i>Myotonia</i>			Yes
<i>Frontal balding</i>			Yes
<i>Classic facies</i>			Yes
<i>Lens involvement</i>			Yes
<i>Cognitive impairment</i>			Yes
<i>Pigmentary retinopathy</i>			Yes
<i>♥ conduction issues</i>			Yes
<i>Onset late teens</i>			Yes

*(list is not exhaustive)*



**Q** *Chronic Progressive External Ophthalmoplegia (CPEO)*

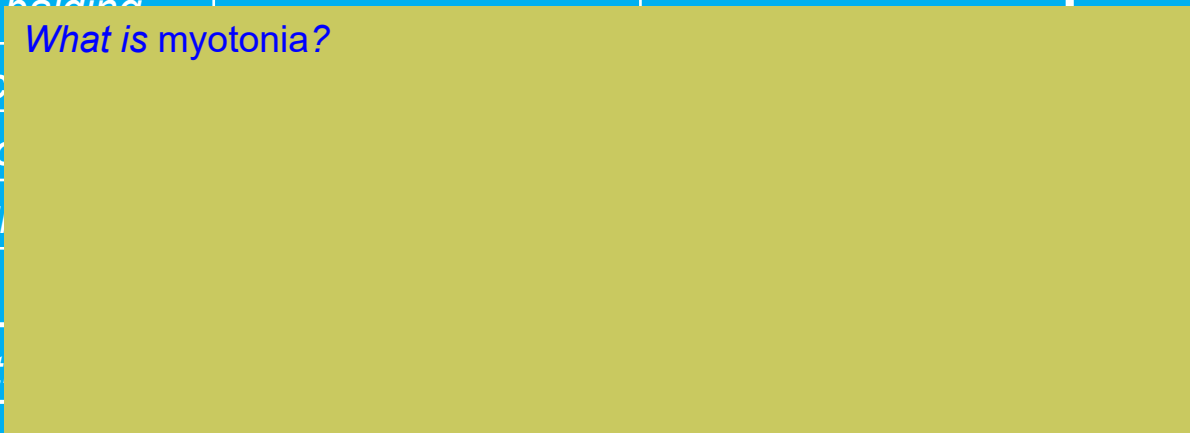
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<i>Clinical course</i>	Waxes and wanes	Progressive	
<i>Diplopia?</i>	Yes	No	
<b>Myotonia</b>			Yes
<i>Frontal holding</i>			Yes
<i>Classic</i>			Yes
<i>Lens invol</i>			Yes
<i>Cognitive i</i>			Yes
<i>Pigmentary</i>			Yes
<i>♥ conduct</i>			Yes
<i>Onset late teens</i>			Yes

What is myotonia?





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Frontal holding			Yes
Classic			Yes
Lens invol			Yes
Cognitive i			Yes
Pigmentary			Yes
♥ conduct			Yes
Onset late teens			Yes

What is myotonia?  
 The inability of a muscle to relax after contraction



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Lens invol			Yes
Cognitive i			Yes
Pigmentary			Yes
♥ conduct			Yes
Onset late teens			Yes

What is myotonia?  
 The inability of a muscle to relax after contraction

What is the classic story for recognizing a pt suffers from myotonia?





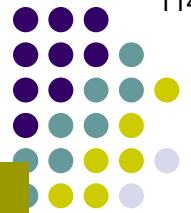
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+Tensilon test?	Yes	No	
Clinical course	Waxes and wanes	Progressive	
Diplopia?	Yes	No	
<b>Myotonia</b>			Yes
Frontal holding			Yes
Classic	The inability of a muscle to relax after contraction		Yes
Lens invol	What is the classic story for recognizing a pt suffers from myotonia?		Yes
Cognitive i	When you greet them in clinic, the handshake is prolonged because they have difficulty letting go		Yes
Pigmentary			Yes
♥ conduct			Yes
Onset late teens			Yes

What is myotonia?  
 The inability of a muscle to relax after contraction

What is the classic story for recognizing a pt suffers from myotonia?  
 When you greet them in clinic, the handshake is prolonged because they have difficulty letting go



# Q Chronic Progressive External Ophthalmoplegia (CPEO)

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Diplopia?	Yes	No	
<b>Myotonia</b>	?	?	Yes
Frontal holding			Yes
Classic	The inability of a muscle to relax after contraction		Yes
Lens invol	What is the classic story for recognizing a pt suffers from myotonia?		Yes
Cognitive i	When you greet them in clinic, the handshake is prolonged because they have difficulty letting go		Yes
Pigmentary			Yes
♥ conduct	Is myotonia a manifestation of MG or CPEO?		Yes
Onset late teens			Yes

What is myotonia?  
 The inability of a muscle to relax after contraction

What is the classic story for recognizing a pt suffers from myotonia?  
 When you greet them in clinic, the handshake is prolonged because they have difficulty letting go

Is myotonia a manifestation of MG or CPEO?



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Clinical course	Waxes and wanes	Progressive	
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♥ conduct			Yes
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What is the classic story for recognizing a pt suffers from myotonia?  
 When you greet them in clinic, the handshake is prolonged because they have difficulty letting go

Is myotonia a manifestation of MG or CPEO?  
 No



**Q** *Chronic Progressive External Ophthalmoplegia (CPEO)*

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*Myotonic dystrophy has a number of manifestations—identify them.*

	<b>MG</b>	<b>CPEO</b>	<i>Myotonic Dystrophy</i>
<i>Fatigable?</i>	Yes	No	
<i>+Tensilon test?</i>	Yes	No	
<i>Clinical course</i>	Waxes and wanes	Progressive	
<i>Diplopia?</i>	Yes	No	
<i>Myotonia</i>	No	No	Yes
<b>Frontal balding</b>			Yes
<i>Classic facies</i>			Yes
<i>Lens involve</i>			Yes
<i>Cognitive imp</i>			Yes
<i>Pigmentary reti</i>			Yes
♥ <i>conduction issues</i>			Yes
<i>Onset late teens</i>			Yes

*To what does the term frontal balding refer?*



# A Chronic Progressive External Ophthalmoplegia (CPEO)

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 Myotonic dystrophy has a number of manifestations—identify them.

	MG	CPEO	Myotonic Dystrophy
Fatigable?	Yes	No	
+Tensilon test?	Yes	No	
Clinical course	Waxes and wanes	Progressive	
Diplopia?	Yes	No	
Myotonia	No	No	Yes
<b>Frontal balding</b>			Yes
Classic facies			Yes
Lens involve			Yes
Cognitive imp			Yes
Pigmentary reti			Yes
♥ conduction issues			Yes
Onset late teens			Yes

To what does the term frontal balding refer?  
 Just what it sounds like—a specific pattern of hair loss

## Chronic Progressive External Ophthalmoplegia (CPEO)



Myotonic dystrophy: Frontal balding

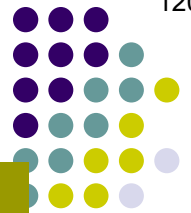


**Q** *Chronic Progressive External Ophthalmoplegia (CPEO)*

*In a nutshell, what sort of condition is myotonic dystrophy?*  
 An inherited ( AD ) progressive systemic condition that results in ophthalmoplegia  
*Myotonic dystrophy has a number of manifestations—identify them.*

	<b>MG</b>	<b>CPEO</b>	<i>Myotonic Dystrophy</i>
<i>Fatigable?</i>	Yes	No	
<i>+Tensilon test?</i>	Yes	No	
<i>Clinical course</i>	Waxes and wanes	Progressive	
<i>Diplopia?</i>	Yes	No	
<i>Myotonia</i>	No	No	Yes
<b>Frontal balding</b>	?	?	Yes
<i>Classic facies</i>			Yes
<i>Lens involve</i>			Yes
<i>Cognitive impo</i>			Yes
<i>Pigmentary reti</i>			Yes
♥ <i>conduction issues</i>			Yes
<i>Onset late teens</i>			Yes

*To what does the term frontal balding refer?*  
 Just what it sounds like—a specific pattern of hair loss  
*Is frontal balding a manifestation of MG or CPEO?*



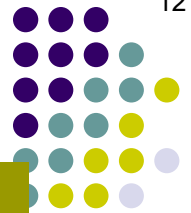
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 Myotonic dystrophy has a number of manifestations—identify them.

	MG	CPEO	Myotonic Dystrophy
Fatigable?	Yes	No	
+Tensilon test?	Yes	No	
Clinical course	Waxes and wanes	Progressive	
Diplopia?	Yes	No	
Myotonia	No	No	Yes
<b>Frontal balding</b>	No	No	Yes
Classic facies			Yes
Lens involve			Yes
Cognitive imp			Yes
Pigmentary reti			Yes
♥ conduction issues			Yes
Onset late teens			Yes

To what does the term frontal balding refer?  
 Just what it sounds like—a specific pattern of hair loss  
 Is frontal balding a manifestation of MG or CPEO?  
 No





**Q** *Chronic Progressive External Ophthalmoplegia (CPEO)*

*In a nutshell, what sort of condition is myotonic dystrophy?*

An inherited ( AD ) progressive systemic condition that results in ophthalmoplegia

*Myotonic dystrophy has a number of manifestations—identify them.*

	<b>MG</b>	<b>CPEO</b>	<b>Myotonic Dystrophy</b>
<i>Fatigable?</i>	Yes	No	
<i>+Tensilon test?</i>	Yes	No	
<i>Clinical course</i>	Waxes and wanes	Progressive	
<i>Diplopia?</i>	Yes	No	
<b>Myotonia</b>	No	No	<b>Yes</b>
<b>Frontal balding</b>	No	No	<b>Yes</b>
<b>Classic facies</b>			<b>Yes</b>
<i>Lens involvement</i>			
<i>Cognitive impairment</i>			
<i>Pigmentary retinopathy</i>			
<i>♥ conduction issues</i>			
<i>Onset late teens</i>			

*By what descriptive term is the classic myotonic-dystrophy facies known?*

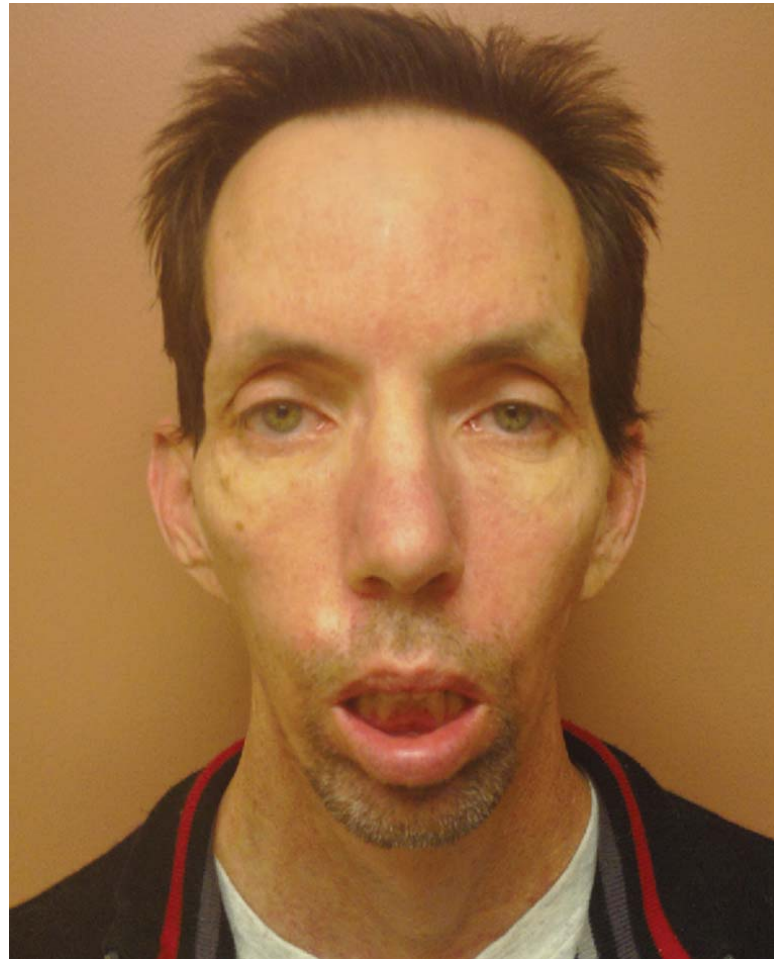
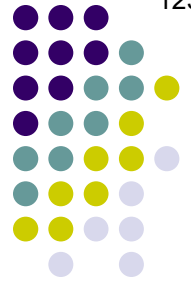


# A Chronic Progressive External Ophthalmoplegia (CPEO)

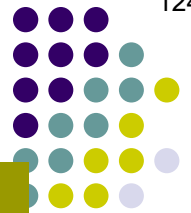
In a nutshell, what sort of condition is myotonic dystrophy?  
 An inherited ( AD ) progressive systemic condition that results in ophthalmoplegia  
 Myotonic dystrophy has a number of manifestations—identify them.

	MG	CPEO	Myotonic Dystrophy
Fatigable?	Yes	No	
+Tensilon test?	Yes	No	
Clinical course	Waxes and wanes	Progressive	
Diplopia?	Yes	No	
Myotonia	No	No	Yes
Frontal balding	No	No	Yes
<b>Classic facies</b>			Yes
Lens involvement	By what descriptive term is the classic myotonic-dystrophy facies known?		
Cognitive impairment	'Hatchet face'		
Pigmentary retinopathy			
♥ conduction issues			
Onset late teens			

*Chronic Progressive External Ophthalmoplegia (CPEO)*



Myotonic dystrophy: 'Hatchet face'



**Q** *Chronic Progressive External Ophthalmoplegia (CPEO)*

*In a nutshell, what sort of condition is myotonic dystrophy?*

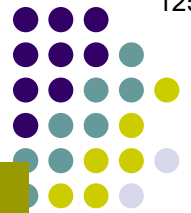
An inherited ( AD ) progressive systemic condition that results in ophthalmoplegia

*Myotonic dystrophy has a number of manifestations—identify them.*

	<b>MG</b>	<b>CPEO</b>	<b>Myotonic Dystrophy</b>
<i>Fatigable?</i>	Yes	No	
<i>+Tensilon test?</i>	Yes	No	
<i>Clinical course</i>	Waxes and wanes	Progressive	
<i>Diplopia?</i>	Yes	No	
<b>Myotonia</b>	No	No	<b>Yes</b>
<b>Frontal balding</b>	No	No	<b>Yes</b>
<b>Classic facies</b>			<b>Yes</b>
<i>Lens involvement</i>			
<i>Cognitive impairment</i>			
<i>Pigmentary retinopathy</i>			
<i>♥ conduction issues</i>			
<i>Onset late teens</i>			

*By what descriptive term is the classic myotonic-dystrophy facies known?  
‘Hatchet face’*

*What changes lead to this appearance?*



# A Chronic Progressive External Ophthalmoplegia (CPEO)

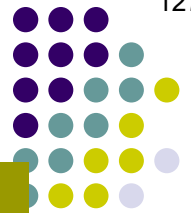
In a nutshell, what sort of condition is myotonic dystrophy?  
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 Myotonic dystrophy has a number of manifestations—identify them.

	MG	CPEO	Myotonic Dystrophy
Fatigable?	Yes	No	
+Tensilon test?	Yes	No	
Clinical course	Waxes and wanes	Progressive	
Diplopia?	Yes	No	
Myotonia	No	No	Yes
Frontal balding	No	No	Yes
<b>Classic facies</b>			Yes
Lens involvement	By what descriptive term is the classic myotonic-dystrophy facies known?		
Cognitive impairment	'Hatchet face'		
Pigmentary retinopathy	What changes lead to this appearance?		
♥ conduction issues	Wasting of the temporalis and masseter muscles		
Onset late teens			

## Chronic Progressive External Ophthalmoplegia (CPEO)



Myotonic dystrophy: 'Hatchet face'—temporalis/masseter wasting



# Q Chronic Progressive External Ophthalmoplegia (CPEO)

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 Myotonic dystrophy has a number of manifestations—identify them.

	MG	CPEO	Myotonic Dystrophy
Fatigable?	Yes	No	
+Tensilon test?	Yes	No	
Clinical course	Waxes and wanes	Progressive	
Diplopia?	Yes	No	
Myotonia	No	No	Yes
Frontal balding	No	No	Yes
<b>Classic facies</b>	?	?	Yes

Lens involvement	<p>By what descriptive term is the classic myotonic-dystrophy facies known?                  'Hatchet face'</p> <p>What changes lead to this appearance?                  Wasting of the temporalis and masseter muscles</p> <p>Is hatchet face a manifestation of MG or CPEO?</p>
Cognitive impairment	
Pigmentary retinopathy	
♥ conduction issues	
Onset late teens	



# A Chronic Progressive External Ophthalmoplegia (CPEO)

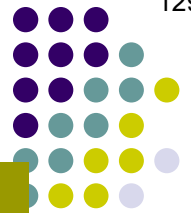
*In a nutshell, what sort of condition is myotonic dystrophy?*

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*Myotonic dystrophy has a number of manifestations—identify them.*

	MG	CPEO	Myotonic Dystrophy
<i>Fatigable?</i>	Yes	No	
<i>+Tensilon test?</i>	Yes	No	
<i>Clinical course</i>	Waxes and wanes	Progressive	
<i>Diplopia?</i>	Yes	No	
<i>Myotonia</i>	No	No	Yes
<i>Frontal balding</i>	No	No	Yes
<b>Classic facies</b>	No	No	Yes
<i>Lens involvement</i>	<i>By what descriptive term is the classic myotonic-dystrophy facies known?</i>		
<i>Cognitive impairment</i>	‘Hatchet face’		
<i>Pigmentary retinopathy</i>	<i>What changes lead to this appearance?</i>		
♥ <i>conduction issues</i>	Wasting of the temporalis and masseter muscles		
<i>Onset late teens</i>	<i>Is hatchet face a manifestation of MG or CPEO?</i>		
	No		





**Q** *Chronic Progressive External Ophthalmoplegia (CPEO)*

*In a nutshell, what sort of condition is myotonic dystrophy?*

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	<b>MG</b>	<b>CPEO</b>	<i>Myotonic Dystrophy</i>
<i>Fatigable?</i>	Yes	No	
<i>+Tensilon test?</i>	Yes	No	
<i>Clinical course</i>	Waxes and wanes	Progressive	
<i>Diplopia?</i>	Yes	No	
<i>Myotonia</i>	No	No	Yes
<i>Frontal balding</i>	No	No	Yes
<i>Classic facies</i>	No	No	Yes
<b>Lens involvement</b>			Yes
<i>Cognitive impairment</i>			✓
<i>Pigmentary retinopathy</i>			✓
<i>♥ conduction issues</i>			✓
<i>Onset late teens</i>			✓

*How is the lens involved in myotonic dystrophy?*



# A Chronic Progressive External Ophthalmoplegia (CPEO)

*In a nutshell, what sort of condition is myotonic dystrophy?*

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	MG	CPEO	Myotonic Dystrophy
<i>Fatigable?</i>	Yes	No	
<i>+Tensilon test?</i>	Yes	No	
<i>Clinical course</i>	Waxes and wanes	Progressive	
<i>Diplopia?</i>	Yes	No	
<i>Myotonia</i>	No	No	Yes
<i>Frontal balding</i>	No	No	Yes
<i>Classic facies</i>	No	No	Yes
<b>Lens involvement</b>			Yes
<i>Cognitive impairment</i>			✓
<i>Pigmentary retinopathy</i>			✓
<i>♥ conduction issues</i>			✓
<i>Onset late teens</i>			✓

*How is the lens involved in myotonic dystrophy?*  
 It is cataractous at an early age, with a unique appearance



**Q** *Chronic Progressive External Ophthalmoplegia (CPEO)*

*In a nutshell, what sort of condition is myotonic dystrophy?*

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*Myotonic dystrophy has a number of manifestations—identify them.*

	MG	CPEO	Myotonic Dystrophy
<i>Fatigable?</i>	Yes	No	
<i>+Tensilon test</i>			
<i>Clinical course</i>			
<i>Diplopia?</i>			
<i>Myotonia</i>			
<i>Frontal balding</i>			
<i>Classic facies</i>	NO	NO	Yes
<b>Lens involvement</b>			Yes
<i>Cognitive impairment</i>			
<i>Pigmentary retinopathy</i>			
<i>♥ conduction issues</i>			
<i>Onset late teens</i>			

*What is the appearance of these cataracts?*

*How is the lens involved in myotonic dystrophy?*  
It is cataractous at an early age **with a unique appearance**



# A Chronic Progressive External Ophthalmoplegia (CPEO)

In a nutshell, what sort of condition is myotonic dystrophy?

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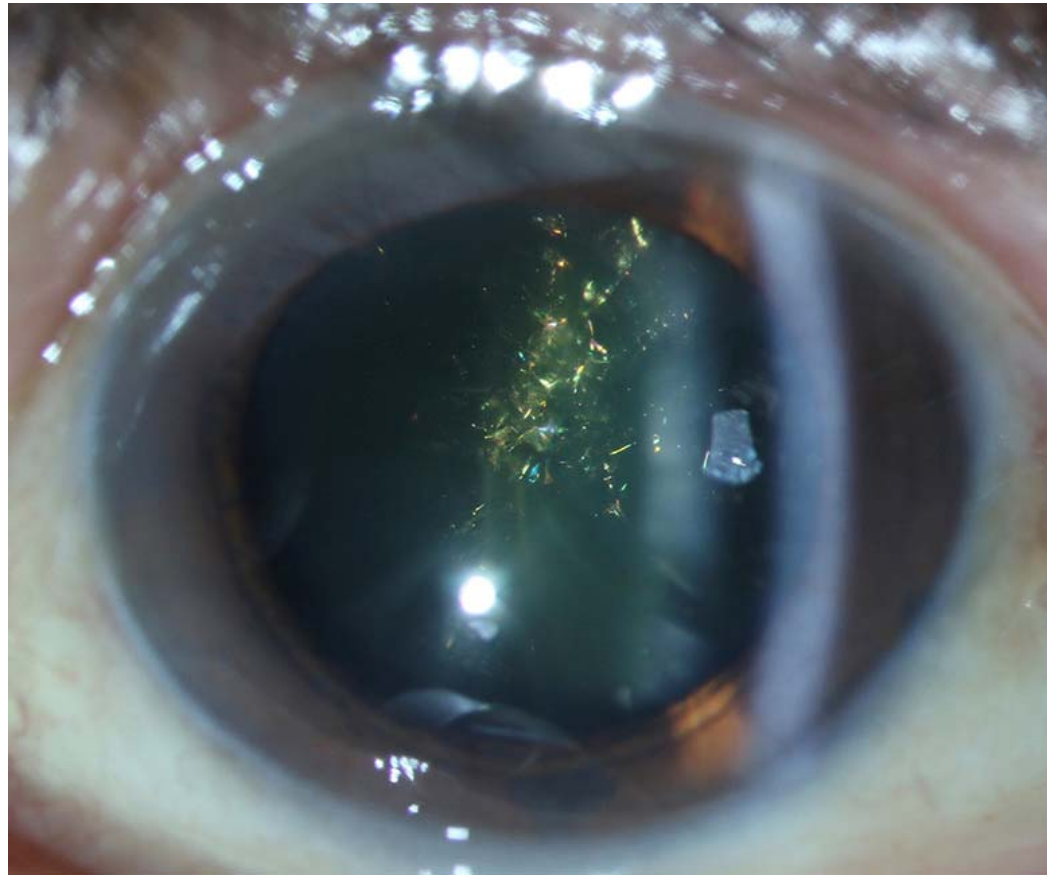
Myotonic dystrophy has a number of manifestations—identify them.

	MG	CPEO	Myotonic Dystrophy
Fatigable?	Yes	No	
+Tensilon test			
Clinical course			
Diplopia?			
Myotonia			
Frontal balding			
Classic facies	NO	NO	Yes
<b>Lens involvement</b>			Yes
Cognitive impairment			
Pigmentary retinopathy			
♥ conduction issues			
Onset late teens			

What is the appearance of these cataracts?  
They appear as iridescent, polychromatic crystal in the lens cortex

How is the lens involved in myotonic dystrophy?  
It is cataractous at an early age **with a unique appearance**

## Chronic Progressive External Ophthalmoplegia (CPEO)



Myotonic dystrophy: Cataract



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 Myotonic dystrophy has a number of manifestations—identify them.

	MG	CPEO	Myotonic Dystrophy
Fatigable?	Yes	No	
+Tensilon test			
Clinical course			
Diplopia?			
Myotonia			
Frontal balding			
Classic facies	NO	NO	Yes
<b>Lens involvement</b>			Yes
Cognitive impairment			
Pigmentary retinopathy			
♥ conduction issues			
Onset late teens			

What is the appearance of these cataracts?  
 They appear as iridescent, polychromatic crystal in the lens cortex  
 By what descriptive term is the classic myotonic-dystrophy cataract known?

How is the lens involved in myotonic dystrophy?  
 It is cataractous at an early age **with a unique appearance**



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	MG	CPEO	Myotonic Dystrophy
Fatigable?	Yes	No	
+Tensilon test			
Clinical course			
Diplopia?			
Myotonia			
Frontal balding			
Classic facies	NO	NO	Yes
<b>Lens involvement</b>			Yes
Cognitive impairment			
Pigmentary retinopathy			
♥ conduction issues			
Onset late teens			

What is the appearance of these cataracts?  
 They appear as iridescent, polychromatic crystal in the lens cortex

By what descriptive term is the classic myotonic-dystrophy cataract known?  
 'Christmas tree cataract'

How is the lens involved in myotonic dystrophy?  
 It is cataractous at an early age with a unique appearance



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<i>Clinical course</i>			
<i>Diplopia?</i>			
<i>Myotonia</i>			
<i>Frontal balding</i>			
<i>Classic facies</i>	NO	NO	Yes
<b>Lens involvement</b>			Yes
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*By what descriptive term is the classic myotonic-dystrophy cataract known?*  
**‘Christmas tree cataract’**

*Are they visually significant?*

*How is the lens involved in myotonic dystrophy?*  
 It is cataractous at an early age **with a unique appearance**





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Fatigable?	Yes	No	
+Tensilon test			
Clinical course	<p>What is the appearance of these cataracts? They appear as iridescent, polychromatic crystal in the lens cortex</p>		
Diplopia?	<p>By what descriptive term is the classic myotonic-dystrophy cataract known? <b>'Christmas tree cataract'</b></p>		
Myotonia	<p>Are they visually significant? Yes—over time, a total cortical cataract will develop, along with a PSC</p>		
Frontal balding			
Classic facies	NO	NO	Yes
<b>Lens involvement</b>			Yes
Cognitive impairment	<p>How is the lens involved in myotonic dystrophy? It is cataractous at an early age <b>with a unique appearance</b></p>		
Pigmentary retinopathy			
♥ conduction issues			
Onset late teens			



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Fatigable?	Yes	No	
+Tensilon test			
Clinical course			
Diplopia?			
Myotonia			
Frontal balding			
Classic facies	NO	NO	Yes
<b>Lens involvement</b>	?	?	Yes
Cognitive impairment			✓
Pigmentary retinopathy			✓
♥ conduction issues			✓
Onset late teens			✓

What is the appearance of these cataracts?  
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By what descriptive term is the classic myotonic-dystrophy cataract known?  
 'Christmas tree cataract'

Are they visually significant?  
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How is the lens involved in myotonic dystrophy?  
 It is cataractous at an early age, with a unique appearance

Are Christmas tree cataracts a manifestation of MG or CPEO?

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Clinical course			
Diplopia?			
Myotonia			
Frontal balding			
Classic facies	NO	NO	Yes
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Cognitive impairment			
Pigmentary retinopathy			
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Onset late teens			

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 No



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<i>Fatigable?</i>	Yes	No	
<i>+Tensilon test?</i>	Yes	No	
<i>Clinical course</i>	Waxes and wanes	Progressive	
<i>Diplopia?</i>	Yes	No	
<i>Myotonia</i>	No	No	Yes
<i>Frontal balding</i>	No	No	Yes
<i>Classic facies</i>	No	No	Yes
<i>Lens involvement</i>	No	No	Yes
<b>Cognitive impairment</b>			Yes
<i>Pigmentary retinopathy</i>	<i>How does cognitive impairment manifest in myotonic dystrophy?</i>		
<i>♥ conduction issues</i>			
<i>Onset late teens</i>			



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<i>+Tensilon test?</i>	Yes	No	
<i>Clinical course</i>	Waxes and wanes	Progressive	
<i>Diplopia?</i>	Yes	No	
<i>Myotonia</i>	No	No	Yes
<i>Frontal balding</i>	No	No	Yes
<i>Classic facies</i>	No	No	Yes
<i>Lens involvement</i>	No	No	Yes
<b>Cognitive impairment</b>			Yes
<i>Pigmentary retinopathy</i>	<p><i>How does cognitive impairment manifest in myotonic dystrophy?</i>                      Pts tend to have low intelligence</p>		
<i>♥ conduction issues</i>			
<i>Onset late teens</i>			



**Q** *Chronic Progressive External Ophthalmoplegia (CPEO)*

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	<b>MG</b>	<b>CPEO</b>	<b>Myotonic Dystrophy</b>
<i>Fatigable?</i>	Yes	No	
<i>+Tensilon test?</i>	Yes	No	
<i>Clinical course</i>	Waxes and wanes	Progressive	
<i>Diplopia?</i>	Yes	No	
<i>Myotonia</i>	No	No	Yes
<i>Frontal balding</i>	No	No	Yes
<i>Classic facies</i>	No	No	Yes
<i>Lens involvement</i>	No	No	Yes
<b>Cognitive impairment</b>	?	?	Yes
<i>Pigmentary retinopathy</i>	<p><i>How does cognitive impairment manifest in myotonic dystrophy?</i>                      Pts tend to have low intelligence</p> <p><i>Is low intelligence a manifestation of MG or CPEO?</i></p>		
<i>♥ conduction issues</i>			
<i>Onset late teens</i>			



# A Chronic Progressive External Ophthalmoplegia (CPEO)

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	MG	CPEO	Myotonic Dystrophy
<i>Fatigable?</i>	Yes	No	
<i>+Tensilon test?</i>	Yes	No	
<i>Clinical course</i>	Waxes and wanes	Progressive	
<i>Diplopia?</i>	Yes	No	
<i>Myotonia</i>	No	No	Yes
<i>Frontal balding</i>	No	No	Yes
<i>Classic facies</i>	No	No	Yes
<i>Lens involvement</i>	No	No	Yes
<i>Cognitive impairment</i>	No	No	Yes
<i>Pigmentary retinopathy</i>	<i>How does cognitive impairment manifest in myotonic dystrophy?</i>		
<i>♥ conduction issues</i>	Pts tend to have low intelligence		
<i>Onset late teens</i>	<i>Is low intelligence a manifestation of MG or CPEO?</i>		
	No		



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<i>Fatigable?</i>	Yes	No	
<i>+Tensilon test?</i>	Yes	No	
<i>Clinical course</i>	Waxes and wanes	Progressive	
<i>Diplopia?</i>	Yes	No	
<i>Myotonia</i>	No	No	Yes
<i>Frontal balding</i>	No	No	Yes
<i>Classic facies</i>	No	No	Yes
<i>Lens involvement</i>	No	No	Yes
<i>Cognitive impairment</i>	No	No	Yes
<i>Pigmentary retinopathy</i>	?	?	Yes
♥ <i>conduction issues</i>			Yes
<i>Onset late teens</i>			Yes

*Is pigmentary retinopathy a manifestation of MG or CPEO?*





# A Chronic Progressive External Ophthalmoplegia (CPEO)

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 Myotonic dystrophy has a number of manifestations—identify them.

	MG	CPEO	Myotonic Dystrophy
Fatigable?	Yes	No	
+Tensilon test?	Yes	No	
Clinical course	Waxes and wanes	Progressive	
Diplopia?	Yes	No	
Myotonia	No	No	Yes
Frontal balding	No	No	Yes
Classic facies	No	No	Yes
Lens involvement	No	No	Yes
Cognitive impairment	No	No	Yes
Pigmentary retinopathy	No	Yes (in KSS)	Yes
♥ conduction issues	Is pigmentary retinopathy a manifestation of MG or CPEO?		Yes
Onset late teens	Yes—it is encountered in the KSS form of CPEO		Yes



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	<b>MG</b>	<b>CPEO</b>	<b>Myotonic Dystrophy</b>
<i>Fatigable?</i>	Yes	No	
<i>+Tensilon test?</i>	Yes	No	
<i>Clinical course</i>	Waxes and wanes	Progressive	
<i>Diplopia?</i>	Yes	No	
<i>Myotonia</i>	No	No	Yes
<i>Frontal balding</i>	No	No	Yes
<i>Classic facies</i>	No	No	Yes
<i>Lens involvement</i>	No	No	Yes
<i>Cognitive impairment</i>			Yes
<i>Pigmentary retinopathy</i>			Yes
<b>♥ conduction issues</b>	?	?	Yes
<i>Onset late teens</i>			Yes

*Are cardiac conduction issues a manifestation of MG or CPEO?*



# A Chronic Progressive External Ophthalmoplegia (CPEO)

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Myotonic dystrophy has a number of manifestations—identify them.

	MG	CPEO	Myotonic Dystrophy
Fatigable?	Yes	No	
+Tensilon test?	Yes	No	
Clinical course	Waxes and wanes	Progressive	
Diplopia?	Yes	No	
Myotonia	No	No	Yes
Frontal balding	No	No	Yes
Classic facies	No	No	Yes
Lens involvement	No	No	Yes
Cognitive impairment			Yes
Pigmentary retinopathy			Yes
♥ conduction issues	No	Yes (in KSS)	Yes
Onset late teens			Yes

Are cardiac conduction issues a manifestation of MG or CPEO?  
 Yes—they are encountered in the KSS form of CPEO



**Q** *Chronic Progressive External Ophthalmoplegia (CPEO)*

*In a nutshell, what sort of condition is myotonic dystrophy?*  
 An inherited ( AD ) progressive systemic condition that results in ophthalmoplegia  
*Myotonic dystrophy has a number of manifestations—identify them.*

	<b>MG</b>	<b>CPEO</b>	<b>Myotonic Dystrophy</b>
<i>Fatigable?</i>	Yes	No	
<i>+Tensilon test?</i>	Yes	No	
<i>Clinical course</i>	Waxes and wanes	Progressive	
<i>Diplopia?</i>	Yes	No	
<i>Myotonia</i>	No	No	Yes
<i>Frontal balding</i>	No	No	Yes
<i>Classic facies</i>	No	No	Yes
<i>Lens involvement</i>	No	No	Yes
<i>Cognitive impairment</i>	No	No	Yes
<i>Pigmentary retinopathy</i>			
<i>♥ conduction issues</i>			
<b>Onset late teens</b>	?	?	Yes

*Is onset during the teen years typical for MG or CPEO?*

**Onset late teens**



# A Chronic Progressive External Ophthalmoplegia (CPEO)

In a nutshell, what sort of condition is myotonic dystrophy?

An inherited ( AD ) progressive systemic condition that results in ophthalmoplegia

Myotonic dystrophy has a number of manifestations—identify them.

	MG	CPEO	Myotonic Dystrophy
Fatigable?	Yes	No	
+Tensilon test?	Yes	No	
Clinical course	Waxes and wanes	Progressive	
Diplopia?	Yes	No	
Myotonia	No	No	Yes
Frontal balding	No	No	Yes
Classic facies	No	No	Yes
Lens involvement	No	No	Yes
Cognitive impairment	No	No	Yes
Pigmentary retinopathy	No	No	Yes
♥ conduction issues	No	Yes (rare)	Yes
<b>Onset late teens</b>	Unusual	Yes	Yes

Is onset during the teen years typical for MG or CPEO?  
 While it would be atypical (but possible) in MG, it would be typical for CPEO

## Chronic Progressive External Ophthalmoplegia (CPEO)

In a nutshell, what sort of condition is myotonic dystrophy?

An inherited ( AD ) progressive systemic condition that results in ophthalmoplegia

Myotonic dystrophy has a number of manifestations—identify them.

	MG	CPEO	Myotonic Dystrophy
<i>Fatigable?</i>	Yes	No	
So upon encountering a young-adult pt with bilateral ptosis, pigmentary retinopathy and cardiac conduction issues, don't jump straight to CPEO...			
<i>Myotonia</i>	No	No	Yes
<i>Frontal balding</i>	No	No	Yes
<i>Classic facies</i>	No	No	Yes
<i>Lens involvement</i>	No	No	Yes
<i>Cognitive impairment</i>	No	No	Yes
<i>Pigmentary retinopathy</i>	No	Yes (in KSS)	Yes
<i>♥ conduction issues</i>	No	Yes (in KSS)	Yes
<i>Onset late teens</i>	Unusual	Yes	Yes

## Chronic Progressive External Ophthalmoplegia (CPEO)

In a nutshell, what sort of condition is myotonic dystrophy?

An inherited ( AD ) progressive systemic condition that results in ophthalmoplegia

Myotonic dystrophy has a number of manifestations—identify them.

	MG	CPEO	Myotonic Dystrophy
Fatigable?	Yes	No	
<p>So upon encountering a young-adult pt with bilateral ptosis, pigmentary retinopathy and cardiac conduction issues, don't jump straight to CPEO...  <i>Be sure to check for signs/symptoms of myotonic dystrophy first!</i></p>			
Myotonia	No	No	Yes
Frontal balding	No	No	Yes
Classic facies	No	No	Yes
Lens involvement	No	No	Yes
Cognitive impairment	No	No	Yes
Pigmentary retinopathy	No	Yes (in KSS)	Yes
♥ conduction issues	No	Yes (in KSS)	Yes
Onset late teens	Unusual	Yes	Yes