Which of the following concerning CPEO are true?

- It begins with ptosis, usually after age 60
Chronic Progressive External Ophthalmoplegia (CPEO)

- Which of the following concerning CPEO are true?
  - It begins with ptosis, usually after age 60  \( F \)
Which of the following concerning CPEO are true?

- It begins with ptosis, usually after age 60. **T**

Chronic Progressive External Ophthalmoplegia (CPEO)
Which of the following concerning CPEO are true?

- It begins with ptosis, usually after age 60. **False**
- It is a mitochondrial disease. **True**
Chronic Progressive External Ophthalmoplegia (CPEO)

- Which of the following concerning CPEO are true?
  - It begins with ptosis, usually after age 60: **F T**
  - It is a mitochondrial disease: **T**
CPEO in childhood

Which of the following concerning CPEO are true?

- It begins with ptosis, usually after age 60 - F T
- It is a mitochondrial disease - T
- Transmission can be maternal, AR, AD, or sporadic
Chronic Progressive External Ophthalmoplegia (CPEO)

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

- It begins with ptosis, usually after age 60. **T**
- It is a mitochondrial disease. **T**
- Transmission can be maternal, AR, AD, or sporadic. **T**

**How does maternal transmission manifest?**
Which of the following concerning CPEO are true?

- It begins with ptosis, usually after age 60: **F**
- It is a mitochondrial disease: **T**
- Transmission can be maternal, AR, AD, or sporadic: **T**

**How does maternal transmission manifest?**
Only mothers can transmit the dz, and both male and female offspring can get it.
Which of the following concerning CPEO are true?

- It begins with ptosis, usually after age 60: **F**
- It is a mitochondrial disease: **T**
- Transmission can be maternal, AR, AD, or sporadic: **T**

Huh? I thought mitochondrial diseases were always transmitted in maternal fashion. How can a mitochondrial (Mt) dz be transmitted AR or AD?
A Chronic Progressive External Ophthalmoplegia (CPEO)

Which of the following concerning CPEO are true?

- It begins with ptosis, usually after age 60  
  - F  
  - T

- It is a mitochondrial disease  
  - T

- Transmission can be maternal, AR, AD, or sporadic  
  - T

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.
Chronic Progressive External Ophthalmoplegia (CPEO)

- Which of the following concerning CPEO are true?
  - It begins with ptosis, usually after age 60 - T
  - It is a mitochondrial disease - T
  - Transmission can be maternal, AR, AD, or sporadic - T

OK, but which form of transmission is most common?
Which of the following concerning CPEO are true?

- It begins with ptosis, usually after age 60  
  - F
- It is a mitochondrial disease  
  - T
- Transmission can be maternal, AR, AD, or sporadic  
  - T

**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.
Which of the following concerning CPEO are true?

- It begins with ptosis, usually after age 60: **F**
- It is a mitochondrial disease: **T**
- Transmission can be maternal, AR, AD, or sporadic: **T**
- It is associated with VF and ERG abnormalities: **T**
Which of the following concerning CPEO are true?

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- Diplopia is a common complaint
Chronic Progressive External Ophthalmoplegia (CPEO)

- Which of the following concerning CPEO are true?
  - It begins with ptosis, usually after age 60: **F**
  - 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 a common complaint: **F**
A Chronic Progressive External Ophthalmoplegia (CPEO)

- Which of the following concerning CPEO are true?
  - It begins with ptosis, usually 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 a common complaint: **F T**
Chronic Progressive External Ophthalmoplegia (CPEO)

Which of the following concerning CPEO are true?

- It begins with ptosis, usually after age 60: **F**
- 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 a common complaint: **F**

Why is diplopia uncommon in CPEO?

**Because the ophthalmoplegia is usually symmetric bilaterally.**
Chronic Progressive External Ophthalmoplegia (CPEO)

- Which of the following concerning CPEO are true?
  - It begins with ptosis, usually 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 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
Chronic Progressive External Ophthalmoplegia (CPEO)

Which of the following concerning CPEO are true?

- It begins with ptosis, usually 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 a common complaint  
  - F
  - T
- It may be associated with cardiac abnormalities
Chronic Progressive External Ophthalmoplegia (CPEO)

- Which of the following concerning CPEO are true?
  - It begins with ptosis, usually after age 60: **F**
  - 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 a common complaint: **F**
  - It may be associated with cardiac abnormalities: **T**
Chronic Progressive External Ophthalmoplegia (CPEO)

CPEO: Complete heart block in a 15 y.o.
Which of the following concerning CPEO are true?

- It begins with ptosis, usually 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 a common complaint  
  [F T]
- It may be associated with cardiac abnormalities  
  [T]
- It may be associated with a pigmentary retinopathy  
  [T]
Which of the following concerning CPEO are true?

- It begins with ptosis, usually after age 60. [F]
- 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 a common complaint. [F]
- It may be associated with cardiac abnormalities. [T]
- It may be associated with a pigmentary retinopathy. [T]
Chronic Progressive External Ophthalmoplegia (CPEO)

CPEO: Pigmentary retinopathy
Chronic Progressive External Ophthalmoplegia (CPEO)

Q: Which of the following concerning CPEO are true?

- It begins with ptosis, usually after age 60: **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 a common complaint: **F**
- It may be associated with cardiac abnormalities: **T**
- It may be associated with a pigmentary retinopathy: **T**

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

Kearns-Sayre syndrome (KSS)

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?

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

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

Kearns-Sayre syndrome (KSS)

- Diplopia is a common complaint
- It may be associated with **cardiac abnormalities**
- It may be associated with a **pigmentary retinopathy**
Which of the following concerning CPEO are true?

- It begins with ptosis, usually after age 60: **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 a common complaint: **F**
- It may be associated with cardiac abnormalities: **T**
- It may be associated with pigmentary retinopathy: **T**

**Q**

*C Chronic Progressive External Ophthalmoplegia (CPEO)*

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

Kearns-Sayre syndrome (KSS)

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

- Diplopia is a common complaint: **T**
- It may be associated with cardiac abnormalities: **T**
- It may be associated with pigmentary retinopathy: **T**
Chronic Progressive External Ophthalmoplegia (CPEO) in childhood is an uncommon syndrome.

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

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)

- Diplopia is a common complaint
- It may be associated with cardiac abnormalities
- It may be associated with a pigmentary retinopathy
**Chronic Progressive External Ophthalmoplegia (CPEO)**

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

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
- It may be associated with **cardiac abnormalities**
- It may be associated with a **pigmentary retinopathy**
CPEO + pigmentary retinopathy + cardiac conduction problems = what syndrome?
Kearns-Sayre syndrome (KSS)

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?
Precede

- Diplopia is a common complaint
- It may be associated with cardiac abnormalities
- It may be associated with a pigmentary retinopathy
Which of the following concerning CPEO are true?

- It begins with ptosis, usually after age 60: **true**
- It is a mitochondrial disease: **true**
- Transmission can be maternal, AR, AD, or sporadic: **true**
- It is associated with VF and ERG abnormalities: **true**
- Diplopia is a common complaint: **false**
- It may be associated with cardiac abnormalities: **true**
- It may be associated with a pigmentary retinopathy: **true**

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**Q**

*Cerebral Progressive External Ophthalmoplegia (CPEO)*

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

Kearns-Sayre syndrome (KSS)

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?

**Precede**

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

What does this imply re management?

It implies that all cases of progressive ptosis + ophthalmoplegia should undergo cardiac evaluation.
Which of the following concerning CPEO are true?

- It begins with ptosis, usually after age 60: **False**
- It is a mitochondrial disease: **True**
- Transmission can be maternal, AR, AD, or sporadic: **True**
- It is associated with VF and ERG abnormalities: **True**
- Diplopia is a common complaint: **False**
- It may be associated with cardiac abnormalities: **True**
- It may be associated with a pigmentary retinopathy: **True**

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

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 the cardiac issues?
Precede

What does this imply re management?
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- Diplopia is a common complaint
- It may be associated with cardiac abnormalities
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Chronic Progressive External Ophthalmoplegia (CPEO)

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

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? 
Precede

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

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

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

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

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? **Precede**

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

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

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
Chronic Progressive External Ophthalmoplegia (CPEO)

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

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).

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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). One is CPEO/KSS. What are the other three?
Chronic Progressive External Ophthalmoplegia (CPEO)

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

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 cardiac issues?
Precede

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

Myotonic dystrophy, CPEO/KSS, LHON, Leigh syndrome

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

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A  

**Chronic Progressive External Ophthalmoplegia (CPEO)**

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

An inherited AD progressive systemic condition that results in ophthalmoplegia

Myotonic dystrophy

CPEO/KSS

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What are its other ocular manifestations?
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**Q/A**

**Chronic Progressive External Ophthalmoplegia (CPEO)**

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

In a nutshell, what sort of condition is myotonic dystrophy?
An inherited AD progressive systemic condition that results in ophthalmoplegia

What are its other ocular manifestations?
-- Bilateral symmetric ptosis
-- Pigmentary retinopathy
-- 'Christmas tree' cataracts

What are its classic nonocular findings?
-- Cardiac conduction issues
-- Myotonia
-- Characteristic 'hatchet' facies
-- Frontal balding
-- Low intelligence

In a nutshell, what sort of condition is myotonic dystrophy?
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**A Chronic Progressive External Ophthalmoplegia (CPEO)**

CPEO + pigmentary retinopathy + cardiac conduction problems = what syndrome?
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Myotonic dystrophy  
CPEO/KSS  
LHON  
Leigh syndrome
CPEO + pigmentary retinopathy + cardiac conduction problems = what syndrome?
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CPEO + pigmentary retinopathy + cardiac conduction problems = what syndrome? Kearns-Sayre syndrome (KSS)

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? Precede

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

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

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

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

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Chronic Progressive External Ophthalmoplegia (CPEO)

LHON: Progression of ONH atrophy
Chronic Progressive External Ophthalmoplegia (CPEO)

LHON: Central/ceocentral scotomata
CPEO + pigmentary retinopathy + cardiac conduction problems = what syndrome?
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**Chronic Progressive External Ophthalmoplegia (CPEO)**

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Myotonic dystrophy  CPEO/KSS  LHON  Leigh syndrome

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Yes, about 90% of pts are male

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

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 the cardiac issues? Precede

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

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

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?
Precede

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

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.) It is a mitochondrial condition that presents in childhood with cognitive and motor decline, ophthalmoplegia, and optic atrophy.

Leigh syndrome

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

Leigh syndrome

Cardiac abnormalities

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?
Which of the following concerning CPEO are true?

- It begins with ptosis, usually after age 60 - \textbf{F}
- It is a mitochondrial disease - \textbf{T}
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- It is associated with VF and ERG abnormalities - \textbf{T}
- Diplopia is a common complaint - \textbf{F}
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- It is associated with VF and FRG abnormalities. **T**
- Diplopia is a common complaint. **F**
- It may be associated with cardiac abnormalities. **T**
- It may be associated with a pigmentary retinopathy. **T**

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

The pupil is not involved. **F**
Chronic Progressive **External Ophthalmoplegia** (CPEO)

- Which of the following concerning CPEO are true?
  - It begins with ptosis, usually 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 a common complaint - **F**
  - It may be associated with cardiac abnormalities - **T**
  - It may be associated with a pigmentary retinopathy - **T**
  - The pupil is involved - **F** **T**

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

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.
Chronic Progressive External Ophthalmoplegia (CPEO)

- Which of the following concerning CPEO are true?
  - It begins with ptosis, usually after age 60  
  - It is a mitochondrial disease  
  - Transmission can be maternal, AR, AD, or sporadic  
  - It is associated with VF and ERG abnormalities  
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  - It may be associated with cardiac abnormalities  
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The pupil is invo
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- The pupil is involved? **F**

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Which of the following concerning CPEO are true?

- It begins with ptosis, usually after age 60? **F**
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- The pupil is involved? **F**
- It is diagnosed via serology? **F**
Chronic Progressive External Ophthalmoplegia (CPEO)

- Which of the following concerning CPEO are true?
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  - It may be associated with a pigmentary retinopathy: **T**
  - The pupil is involved: **F**
  - It is diagnosed via serology: **F**

- In childhood, CHRONIC PROGRESSIVE EXTERNAL OPHTHALMOPLEGIA IS AN UNCOMMON DISEASE.
Which of the following concerning CPEO are true?

- It begins with ptosis, usually after age 60: F
- 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 a common complaint: F
- It may be associated with cardiac abnormalities: T
- It may be associated with a pigmentary retinopathy: T
- The pupil is involved: T
- It is diagnosed via muscle biopsy or muscle biopsy: F
- It is diagnosed via serology: F
Chronic Progressive External Ophthalmoplegia (CPEO)

Q Which of the following concerning CPEO are true?

- It begins with ptosis, usually after age 60
  - F
- 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 a common complaint
  - F
- It may be associated with cardiac abnormalities
  - T
- It may be associated with a pigmentary retinopathy
  - T
- The pupil is involved
  - F
- It is diagnosed via serology
  - F

What does muscle biopsy reveal?

muscle biopsy
Which of the following concerning CPEO are true?

- It begins with ptosis, usually 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 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: F (T)
- It is diagnosed via serology: F (T)

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’
Which of the following concerning CPEO are true?

- It begins with ptosis, usually after age 60: **F**
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- The pupil is involved: **F**
- Myasthenia gravis (MG) is in the DDx for CPEO: **F**
Chronic Progressive External Ophthalmoplegia (CPEO)

- Which of the following concerning CPEO are true?
  - It begins with ptosis, usually after age 60: **False**
  - It is a mitochondrial disease: **True**
  - Transmission can be maternal, AR, AD, or sporadic: **True**
  - It is associated with VF and ERG abnormalities: **True**
  - Diplopia is a common complaint: **False**
  - It may be associated with cardiac abnormalities: **True**
  - It may be associated with a pigmentary retinopathy: **True**
  - The pupil is involved: **False**
  - It is diagnosed via muscle biopsy: **False**
  - It is diagnosed via serology: **False**
  - Myasthenia gravis (MG) is in the DDx for CPEO: **True**
Chronic Progressive External Ophthalmoplegia (CPEO)

- Which of the following concerning CPEO are true?
  - It begins with ptosis, usually after age 60: **T**
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  - It may be associated with cardiac abnormalities: **T**
  - It may be associated with a pigmentary retinopathy: **T**
  - The pupil is involved: **F**
  - It is diagnosed via serology: **F**
  - Myasthenia gravis (MG) is in the DDx for CPEO: **T**

**Being able to differentiate CPEO from MG is important, so let’s drill down on this**
Which of the following concerning CPEO are true?

- It begins with ptosis, usually after age 60 — False (F)
- It is a mitochondrial disease — True (T)
- Transmission can be maternal, AR, AD, or sporadic — True (T)
- It is associated with VF and ERG abnormalities — True (T)
- Diplopia is a common complaint — False (F)
- It may be associated with cardiac abnormalities — True (T)
- It may be associated with a pigmentary retinopathy — True (T)
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- Diplopia is a common complaint [F]
- It may be associated with cardiac abnormalities [T]
- It may be associated with a pigmentary retinopathy [T]
- The pupil is involved [F, T]
- It is diagnosed via muscle biopsy [not muscle biopsy, F, T]
- It is diagnosed via serology [F, T]
- Myasthenia gravis (MG) is in the DDx for CPEO [T]

What does it mean to say MG is fatigable?

Fatigable? | MG | CPEO
---|---|---
Yes | | No

Fatigable means (with regard to ptosis) that sustained lid elevation will lead to further weakening (i.e., will fatigue) of the levator, causing the ptosis to worsen.
**Chronic Progressive External Ophthalmoplegia (CPEO)**

- Which of the following concerning CPEO are true?
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  - It may be associated with a pigmentary retinopathy
    - **T**
  - The pupil is involved
    - **F**
  - It is diagnosed via serology
    - **F**
  - Myasthenia gravis (MG) is in the DDx for CPEO
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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.
Chronic Progressive External Ophthalmoplegia (CPEO)

MG: Fatigability

In primary  After sustained upgaze  After further sustained upgaze
Which of the following concerning CPEO are true?

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  - **F**

- 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 a common complaint
  - **F**

- It may be associated with cardiac abnormalities
  - **T**

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

- The pupil is not involved
  - **F**

- The pupil is involved
  - **T**

- It is diagnosed via serology
  - **F**

- It is diagnosed via muscle biopsy
  - **F**

- Myasthenia gravis (MG) is in the DDx for CPEO
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### Chronic Progressive External Ophthalmoplegia (CPEO)

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**What is the trade name for Tensilon?**

Edrophonium (so this is aka the edrophonium test)
Which of the following concerning CPEO are true?

- It begins with ptosis, usually after age 60: **T**
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- It is diagnosed via serology: **F**
- Myasthenia gravis (MG) is in the DDx for CPEO: **T**

### Table: CPEO vs. MG

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**Chronic Progressive External Ophthalmoplegia (CPEO)**

in childhood
Chronic Progressive External Ophthalmoplegia (CPEO)

- Which of the following concerning CPEO are true?
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**What is the trade name for Tensilon?**
Edrophonium (so this is aka the *edrophonium test*)

**Pharmacologically, what is edrophonium?**

- Short (quite)
Chronic Progressive External Ophthalmoplegia (CPEO)

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

Pharmacologically, what is edrophonium? An acetylcholinesterase inhibitor
Chronic Progressive External Ophthalmoplegia (CPEO)

Which of the following concerning CPEO are true?

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Pharmacologically, what is edrophonium? An acetylcholinesterase inhibitor

Is it short-, or long-acting? Short (quite)
Chronic Progressive External Ophthalmoplegia (CPEO)

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- It may be associated with a pigmentary retinopathy  
  - T  
- The pupil is involved  
  - F  
- It is diagnosed via serology  
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- Myasthenia gravis (MG) is in the DDx for CPEO  
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### CPEO vs MG

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**What is the trade name for Tensilon?**

Edrophonium (so this is aka the edrophonium test)

**Pharmacologically, what is edrophonium?**

An acetylcholinesterase inhibitor

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

Short (quite)

**What constitutes a positive test?**

It ameliorates them (temporarily)
Which of the following concerning CPEO are true?

- It begins with ptosis, usually after age 60
- It is a mitochondrial disease
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<th>CPEO</th>
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What is the trade name for Tensilon? Edrophonium (so this is aka the edrophonium test)

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
Chronic Progressive External Ophthalmoplegia (CPEO)

MG: Tensilon test

Pre-injection

A few seconds post-injection

MG: Tensilon test
Which of the following concerning CPEO are true?

- It begins with ptosis, usually after age 60
- It is a mitochondrial disease
- Transmission can be maternal, AR, AD, or sporadic
- It is associated with VF and ERG abnormalities
- Diplopia is a common complaint
- It may be associated with cardiac abnormalities
- It may be associated with a pigmentary retinopathy
- The pupil is involved
- It is diagnosed via serology
- Myasthenia gravis (MG) is in the DDx for CPEO

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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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- The temporary amelioration of MG S/S
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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.
Which of the following concerning CPEO are true?

- It begins with ptosis, usually after age 60: **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 a common complaint: **F**
- It may be associated with cardiac abnormalities: **T**
- It may be associated with a pigmentary retinopathy: **T**
- The pupil is not involved: **F**
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<td>Clinical course</td>
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Chronic Progressive External Ophthalmoplegia (CPEO)
Chronic Progressive External Ophthalmoplegia (CPEO)

Which of the following concerning CPEO are true?

- It begins with ptosis, usually after age 60. **F**
- 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 a common complaint. **F**
- It may be associated with cardiac abnormalities. **T**
- It may be associated with a pigmentary retinopathy. **T**
- The pupil is involved. **F**
- It is diagnosed via serology. **F**
- Myasthenia gravis (MG) is in the DDx for CPEO. **T**

### Comparison Table

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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.**
Chronic Progressive External Ophthalmoplegia (CPEO)

CPEO: Progressive ptosis
**Which of the following concerning CPEO are true?**

- It begins with ptosis, usually after age 60—**F**
- 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 a common complaint—**F**
- It may be associated with cardiac abnormalities—**T**
- It may be associated with a pigmentary retinopathy—**T**
- The pupil is involved—**F**
- It is diagnosed via muscle biopsy—**T**
- It is diagnosed via serology—**F**
- Myasthenia gravis (MG) is in the DDx for CPEO—**T**

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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.

<table>
<thead>
<tr>
<th></th>
<th>MG</th>
<th>CPEO</th>
<th>TED</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Fatigable?</strong></td>
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<td>No</td>
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<td>Yes</td>
<td>No</td>
<td>No</td>
</tr>
<tr>
<td><strong>Clinical course</strong></td>
<td>Waxes and wanes</td>
<td>Progressive</td>
<td>Variable—may progress, or resolve</td>
</tr>
<tr>
<td><strong>Diplopia?</strong></td>
<td>Yes</td>
<td>No</td>
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- It may be associated with cardiac abnormalities  **T**
- It may be associated with a pigmentary retinopathy  **T**
- The pupil is **not** involved  **T**
- The pupil is **involved**  **T**
- It is diagnosed via serology  **F**
- It is diagnosed via muscle biopsy  **F**
- Myasthenia gravis (MG) is in the DDx for CPEO  **T**
Chronic Progressive External Ophthalmoplegia (CPEO)

Which of the following concerning CPEO are true?

- It begins with ptosis, usually after age 60: \(\text{T}\)
- It is a mitochondrial disease: \(\text{T}\)
- Transmission can be maternal, AR, AD, or sporadic: \(\text{T}\)
- It is associated with VF and ERG abnormalities: \(\text{T}\)
- Diplopia is a common complaint: \(\text{F}\)
- It may be associated with cardiac abnormalities: \(\text{T}\)
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- It is diagnosed via serology: \(\text{F}\)
- Myasthenia gravis (MG) is in the DDx for CPEO: \(\text{T}\)

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?

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It may be associated with cardiac abnormalities: \(\text{T}\)

It may be associated with a pigmentary retinopathy: \(\text{T}\)

The pupil is involved: \(\text{F}\)

Muscle biopsy: \(\text{F}\)

It is diagnosed via serology: \(\text{F}\)

Myasthenia gravis (MG) is in the DDx for CPEO: \(\text{T}\)
Which of the following concerning CPEO are true?

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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?

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Chronic Progressive External Ophthalmoplegia (CPEO)

Myotonic dystrophy
Chronic Progressive External Ophthalmoplegia (CPEO)

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

- It begins in adults, usually after age 60
- It is a mitochondrial disease
- Transmission can be maternal, AD, or sporadic
- It is associated with VF and ERG abnormalities
- Diplopia is not a common complaint
- It may be associated with cardiac abnormalities
- It may be associated with a pigmentary retinopathy
- The pupil is involved
- It is not diagnosed via muscle biopsy
- It is diagnosed via serology
- Myasthenia gravis (MG) is in the DDx for CPEO

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In a nutshell, what sort of condition is myotonic dystrophy?
An inherited (AD) progressive systemic condition that results in ophthalmoplegia.

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- It may be associated with cardiac abnormalities T
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**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.*

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<td>Yes</td>
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</tr>
<tr>
<td></td>
<td>Yes</td>
<td>Yes (in KSS)</td>
</tr>
<tr>
<td></td>
<td>Yes</td>
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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.

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</tr>
<tr>
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<td></td>
<td></td>
<td>Yes</td>
</tr>
<tr>
<td>Pigmentary retinopathy</td>
<td></td>
<td></td>
<td>Yes</td>
</tr>
<tr>
<td>♥ conduction issues</td>
<td></td>
<td></td>
<td>Yes</td>
</tr>
<tr>
<td>Onset late teens</td>
<td></td>
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(list is not exhaustive)
**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.**

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**Myotonia**

*What is myotonia?*

- The inability of a muscle to relax after contraction
- 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
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.

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

Myotonia

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? No.
**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.

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**Myotonia**

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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*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.

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

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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?**
Yes.
**Chronic Progressive External Ophthalmoplegia (CPEO)**

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Chronic Progressive External Ophthalmoplegia (CPEO)

Myotonic dystrophy: Frontal balding
**Chronic Progressive External Ophthalmoplegia (CPEO)**

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**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.*

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By what descriptive term is the classic myotonic-dystrophy facies known? ‘Hatchet face’
Chronic Progressive External Ophthalmoplegia (CPEO)

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

What changes lead to this appearance?
Wasting of the temporalis and masseter muscles
Chronic Progressive External Ophthalmoplegia (CPEO)

Myotonic dystrophy: ‘Hatchet face’—temporalis/masseter wasting
**Chronic Progressive External Ophthalmoplegia (CPEO)**

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**Chronic Progressive External Ophthalmoplegia (CPEO)**

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**How does cognitive impairment manifest in myotonic dystrophy?**
Pts tend to have low intelligence

**Is low intelligence a manifestation of MG or CPEO?**
No
**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.

| Fatigable? | MG | Yes | CPEO | No |
| +Tensilon test? | Yes | Yes | No |
| Clinical course | Waxes and wanes | Progressive |
| Diplopia? | Yes | No |
| Myotonia | No | No |
| Frontal balding | No | No |
| Classic facies | No | No |
| Lens involvement | No | No |
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| Lens involvement | No | No |
| Cognitive impairment | No | No |
| Pigmentary retinopathy | Yes (in KSS) | Yes (in KSS) |
| Conduction issues | Yes | Yes |

Is pigmentary retinopathy a manifestation of MG or CPEO?

Yes
### Chronic Progressive External Ophthalmoplegia (CPEO)

**In a nutshell, what sort of condition is myotonic dystrophy?**
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**Is pigmentary retinopathy a manifestation of MG or CPEO?**
Yes—it is encountered in the KSS form of CPEO

**Onset late teens**
Yes
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.

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**Are cardiac conduction issues a manifestation of MG or CPEO?**
*Yes—they are encountered in the KSS form of CPEO*
### Chronic Progressive External Ophthalmoplegia (CPEO)

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**Onset late teens**

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

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*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.

**Onset late teens**  
Unusual  
Yes
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So upon encountering a young-adult pt with bilateral ptosis, pigmentary retinopathy and cardiac conduction issues, don’t jump straight to CPEO…

Be sure to check for signs/symptoms of myotonic dystrophy first!

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.

Myotonia

Frontal balding

Classic facies

Lens involvement

Cognitive impairment

Pigmentary retinopathy

Conduction issues

Onset late teens