Pediatric Iris Cysts

Basic clinical distinction

?  ?
Pediatric Iris Cysts

Basic clinical distinction

Primary

Secondary
Pediatric Iris Cysts

Primary

Secondary

Basic anatomic distinction
Pediatric Iris Cysts

Primary

Epithelial

Stromal

Secondary

Pediatric Iris Cysts

Basic anatomic distinction
Pediatric Iris Cysts

Primary

Epithelial

Secondary

Stromal

Advanced anatomic distinction
Pediatric Iris Cysts

Primary

Epithelial
- Pupillary cysts

Stromal
- Iris pigment epithelium cysts

Secondary

Pediatric Iris Cysts

Advanced anatomic distinction
Pediatric Iris Cysts

Primary

Epithelial

Pupillary cysts

Iris pigment epithelium cysts

Secondary

Stromal

Pediatric Iris Cysts

Pupillary cysts

--Pigment epi cysts at the...[location]
Pediatric Iris Cysts

Primary
- Epithelial
  - Pupillary cysts
  - Iris pigment epithelium cysts

Secondary
- Stromal

Pupillary cysts
-- Pigment epi cysts at the pupil border
Pediatric Iris Cysts

Primary

Epithelial

Pupillary cysts

Secondary

Stromal

Iris pigment epithelium cysts

Pupillary cysts
--Pigment epi cysts at the…pupil border
--Presents in...[age]
Pediatric Iris Cysts

- Primary
  - Epithelial
    - Pupillary cysts
    - Iris pigment epithelium cysts
  - Stromal
- Secondary

Pupillary cysts
--Pigment epi cysts at the pupil border
--Presents in infancy
Pediatric Iris Cysts

Primary

Epithelial

Pupillary cysts
--Pigment epi cysts at the pupil border
--Presents in infancy

Iris pigment epithelium cysts
--2o to separation of...

Secondary

Stromal
Pediatric Iris Cysts

Primary

Epithelial

Iris pigment epithelium cysts

Secondary

Stromal

Pupillary cysts

Pigment epi cysts at the pupil border
Presents in infancy

Iris pigment epithelium cysts
2° to separation of epithelial layers
Pediatric Iris Cysts

Primary

Epithelial

Iris pigment epithelium cysts

Secondary

Stromal

Pediatric Iris Cysts

Pupillary cysts

--Pigment epi cysts at the pupil border
--Presents in infancy

Iris pigment epithelium cysts

--2° to separation of epithelial layers
--Presents in [age range]
Pediatric Iris Cysts

Primary

Epithelial

Pupillary cysts

Iris pigment epithelium cysts

Secondary

Stromal

Pupillary cysts

--Pigment epi cysts at the...pupil border
--Presents in...infancy

Iris pigment epithelium cysts

--2° to separation of...epithelial layers
--Presents in...teens
Pediatric Iris Cysts

Primary

Epithelial

Iris pigment epithelium cysts

Pupillary cysts
--Pigment epi cysts at the pupil border
--Presents in infancy

Secondary

Stromal

Iris pigment epithelium cysts
--2° to separation of epithelial layers
--Presents in teens
--Clinically significant? [Y/N]
Pediatric Iris Cysts

Primary
- Epithelial
  - Pupillary cysts
  - Iris pigment epithelium cysts

Secondary
- Stromal

Pupillary cysts
- Pigment epi cysts at the pupil border
- Presents in infancy

Iris pigment epithelium cysts
- 2° to separation of epithelial layers
- Presents in teens
- Clinically significant? No
Pediatric Iris Cysts

Primary

Epithelial

Iris pigment epithelium cysts

Pupillary cysts
--Pigment epi cysts at the pupil border
--Presents in infancy

Stromal

Pupillary cysts
--Due to stromal sequestration of tissue during embryogenesis

Secondary

Iris pigment epithelium cysts
--2o to separation of epithelial layers
--Presents in teens
--Clinically significant? No
Pediatric Iris Cysts

Primary

Epithelial

Pupillary cysts

Iris pigment epithelium cysts

Secondary

Stromal

Stromal cysts

--Due to stromal sequestration of...epithelial tissue during embryogenesis

Iris pigment epithelium cysts

--2o to separation of...epithelial layers
--Present in...teens
--Clinically significant? No

Pupillary cysts

--Pigment epi cysts at the...pupil border
--Present in...infancy
Pediatric Iris Cysts

Primary

Epithelial

Pupillary cysts
--Pigment epi cysts at the pupil border
--Presents in infancy

Iris pigment epithelium cysts
--2o to separation of epithelial layers
--Presents in teens
--Clinically significant? No

Secondary

Stromal

Stromal cysts
--Due to stromal sequestration of epithelial tissue during embryogenesis
--Presents in...[age]
Pediatric Iris Cysts

Primary

Epithelial

Pupillary cysts

Iris pigment epithelium cysts

Stromal

Stromal cysts

--Due to stromal sequestration of…epithelial tissue during embryogenesis
--Presents in…infancy

Secondary

Iris pigment epithelium cysts

--2o to separation of…epithelial layers
--Presents in…teens
--Clinically significant? No

Pupillary cysts

--Pigment epi cysts at the…pupil border
--Presents in…infancy
Pediatric Iris Cysts

Primary

Stromal

Epithelial

Pupillary cysts

Iris pigment epithelium cysts

Iris pigment epithelium cysts

Stromal cysts
--Due to stromal sequestration of epithelial tissue during embryogenesis
--Present in infancy
--Cysts contain epithelial cell type, may cause enlargement

Pupillary cysts
--Pigment epi cysts at the pupil border
--Presents in infancy

Iris pigment epithelium cysts
--2o to separation of epithelial layers
--Presents in teens
--Clinically significant? No
Pediatric Iris Cysts

Primary

- Stromal cysts
  -- Due to stromal sequestration of epithelial tissue during embryogenesis
  -- Presents in infancy
  -- Cysts contain goblet cells, may cause enlargement

- Epithelial
  - Pupillary cysts
    -- Pigment epi cysts at the pupil border
    -- Presents in infancy
  - Iris pigment epithelium cysts
    -- Secondary to separation of epithelial layers
    -- Presents in teens
    -- Clinically significant? No

Secondary

Stromal cysts
-- Due to stromal sequestration of epithelial tissue during embryogenesis
-- Presents in infancy
-- Cysts contain goblet cells, may cause enlargement
Pediatric Iris Cysts

Primary

Epithelial

Pupillary cysts

--Pigment epi cysts at the pupil border
--Presents in infancy

Iris pigment epithelium cysts

Secondary

Stromal

Stromal cysts

--Due to stromal sequestration of epithelial tissue during embryogenesis
--Presents in infancy
--Cysts contain goblet cells, may cause enlargement

Why does the presence of goblet cells predispose cysts to enlargement?

Pupillary cysts

--Pigment epi cysts at the pupil border
--Presents in infancy

Iris pigment epithelium cysts

--Due to separation of epithelial layers
--Presents in teens
--Clinically significant? No
Pediatric Iris Cysts

Primary

Epithelial

Pupillary cysts

Iris pigment epithelium cysts

Secondary

Stromal

Stromal cysts

--Due to stromal sequestration of...epithelial tissue during embryogenesis

--Presents in...infancy

--Cysts contain...goblet cells, may cause enlargement

Why does the presence of goblet cells predispose cysts to enlargement?
Recall that goblet cells produce...mucin. The production (and sequestration) of...mucin that is responsible for cyst enlargement.
**Pediatric Iris Cysts**

**Primary Iris Cysts**
- **Stromal Cysts**
  - Due to stromal sequestration of epithelial tissue during embryogenesis
  - Presents in infancy
  - Cysts contain goblet cells, may cause enlargement

**Secondary Iris Cysts**
- **Iris Pigment Epithelium Cysts**
  - 2nd to sequestered epithelium
  - Presents in teens
  - Clinically significant? No

**Pupillary Cysts**
- Pigment epi cysts at the pupil border
- Presents in infancy

**Why does the presence of goblet cells predispose cysts to enlargement?**
Recall that goblet cells produce mucin. The production (and sequestration) of mucin that is responsible for cyst enlargement.
Pediatric Iris Cysts

Primary

- Stromal
  - Stromal cysts
    - Due to stromal sequestration of epithelial tissue during embryogenesis
    - Presents in infancy
    - Cysts contain goblet cells, may cause enlargement
    - Result in a number of problems:
      - Obstruct...
      - Secondary...
      - Induce...
      - Cyst leakage

Secondary

- Iris pigment epithelium cysts
  - Pupillary cysts
    - Pigment epi cysts at the pupil border
    - Presents in infancy

Pupillary cysts

- Iris pigment epithelium cysts
  - Pupillary cysts
    - 2o to separation of epithelial layers
    - Presents in teens
    - Clinically significant? No
Pediatric Iris Cysts

Primary

Epithelial

Iris pigment epithelium cysts

Pupillary cysts

--Pigment epi cysts at the pupil border
--Presents in infancy

Iris pigment epithelium cysts

--2° to separation of epithelial layers
--Presents in teens
--Clinically significant? No

Stromal

Stromal cysts

--Due to stromal sequestration of epithelial tissue during embryogenesis
--Presents in infancy
--Cysts contain goblet cells, may cause enlargement
--Result in a number of problems:
  --Obstruct visual axis

Secondary

Iris pigment epithelium cysts

--2° to separation of epithelial layers
--Presents in teens
--Clinically significant? Yes

Pediatric Iris Cysts
Pediatric Iris Cysts

Primary

Epithelial

Stromal

Secondary

Pupillary cysts

-- Pigment epi cysts at the pupil border
-- Presents in infancy

Iris pigment epithelium cysts

-- 2° to separation of epithelial layers
-- Presents in teens
-- Clinically significant? No

Iris pigment epithelium cysts

-- Due to stromal sequestration of epithelial tissue during embryogenesis
-- Presents in infancy
-- Cysts contain goblet cells, may cause enlargement
-- Result in a number of problems:
  -- Obstruct visual axis

Stromal cysts

Are such cysts potentially amblyogenic?
Indeed they are
Pediatric Iris Cysts

Primary
  - Stromal
    - Stromal cysts
      -- Due to stromal sequestration of epithelial tissue during embryogenesis
      -- Presents in infancy
      -- Cysts contain goblet cells, may cause enlargement
      -- Result in a number of problems:
        - Obstruct visual axis
  - Epithelial
    - Pupillary cysts
      -- Pigment epi cysts at the pupil border
      -- Presents in infancy
    - Iris pigment epithelium cysts
      -- 2º to separation of epithelial layers
      -- Presents in teens
      -- Clinically significant? No

Secondary

Are such cysts potentially amblyogenic? Indeed they are
Pediatric Iris Cysts

Primary

Epithelial

Pupillary cysts
--Pigment epi cysts at the pupil border
--Presents in infancy

Iris pigment epithelium cysts
--2o to separation of epithelial layers
--Presents in teens
--Clinically significant? No

Stromal

Iris pigment epithelium cysts
--Due to stromal sequestration of epithelial tissue during embryogenesis
--Presents in infancy
--Cysts contain goblet cells, may cause enlargement
--Result in a number of problems:
   --Obstruct visual axis
   --Secondary...?
Pediatric Iris Cysts

Primary

Epithelial

Pupillary cysts

--Pigment epi cysts at the pupil border
--Presents in infancy

Iris pigment epithelium cysts

--2nd to separation of epithelial layers
--Presents in teens
--Clinically significant? No

Stromal

Stromal cysts

--Due to stromal sequestration of epithelial tissue during embryogenesis
--Presents in infancy
--Cysts contain goblet cells, may cause enlargement
--Result in a number of problems:
  --Obstruct visual axis
  --Secondary glaucoma

Secondary

Iris pigment epithelium cysts

--2nd to separation of...epithelial layers
--Presents in...teens
--Clinically significant? No
Pediatric Iris Cysts

**Primary Iris Cysts**
- Stromal cysts
  - Due to stromal sequestration of epithelial tissue during embryogenesis
  - Presents in infancy
  - Cysts contain goblet cells, may cause enlargement
  - Result in a number of problems:
    - Obstruct visual axis
    - Secondary glaucoma
    - Induce? Cyst leakage → chronic iritis

**Secondary Iris Cysts**
- Iris pigment epithelium cysts
  - Due to separation of epithelial layers
  - Presents in infancy
  - Clinically significant? No

---

**Iris pigment epithelium cysts**
- Pigment epi cysts at the pupil border
- Presents in infancy

**Pupillary cysts**
- Pupil border
- Presents in infancy

---

**Stromal cysts**
- Due to stromal sequestration of epithelial tissue during embryogenesis
- Presents in infancy
- Cysts contain goblet cells, may cause enlargement
- Result in a number of problems:
  - Obstruct visual axis
  - Secondary glaucoma
  - Induce?
Pediatric Iris Cysts

Primary

Epithelial

Iris pigment epithelium cysts

Pupillary cysts

--Pigment epi cysts at the pupil border
--Presents in infancy

Iris pigment epithelium cysts

--2° to separation of epithelial layers
--Presents in teens
--Clinically significant? No

Stromal

Stromal cysts

--Due to stromal sequestration of epithelial tissue during embryogenesis
--Presents in infancy
--Cysts contain goblet cells, may cause enlargement
--Result in a number of problems:
  --Obstruct visual axis
  --Secondary glaucoma
  --Induce corneal decompensation

Secondary

Pediatric Iris Cysts

Iris pigment epithelium cysts
Pediatric Iris Cysts

Primary

Epithelial

Stromal

Secondary

Pupillary cysts
--Pigment epi cysts at the...pupil border
--Presents in...infancy

Iris pigment epithelium cysts
--2° to separation of...epithelial layers
--Presents in...teens
--Clinically significant? No

Stromal cysts
--Due to stromal sequestration of...epithelial tissue during embryogenesis
--Presents in...infancy
--Cysts contain...goblet cells, may cause enlargement
--Result in a number of problems:
  --Obstruct...visual axis
  --Secondary...glaucoma
  --Induce...corneal decompensation
  --Cyst leakage → ?
Pediatric Iris Cysts

Primary

Epithelial

Pupillary cysts

Iris pigment epithelium cysts

Secondary

Stromal

Stromal cysts

--Due to stromal sequestration of...epithelial tissue during embryogenesis
--Presents in...infancy
--Cysts contain...goblet cells, may cause enlargement
--Result in a number of problems:
  --Obstruct...visual axis
  --Secondary...glaucoma
  --Induce...corneal decompensation
  --Cyst leakage \(\rightarrow\) chronic iritis

Pupillary cysts

--Pigment epi cysts at the...pupil border
--Presents in...infancy

Iris pigment epithelium cysts

--2\textdegree to separation of...epithelial layers
--Presents in...teens
--Clinically significant? No
Pediatric Iris Cysts

Primary

Epithelial

Pupillary cysts

-- Pigment epi cysts at the pupil border
-- Presents in infancy

Iris pigment epithelium cysts

-- 2nd to separation of epithelial layers
-- Presents in teens
-- Clinically significant? No

Stromal

Secondary

Pediatric Iris Cysts

Iris pigment epithelium cysts

-- Due to stromal sequestration of epithelial tissue during embryogenesis
-- Presents in infancy
-- Cysts contain goblet cells, may cause enlargement
-- Result in a number of problems:
  -- Obstruct visual axis
  -- Secondary glaucoma
  -- Induce corneal decompensation
  -- Cyst leakage → chronic iritis

Secondary Iris pigment epithelium cysts

-- Due to separation of epithelial layers
-- Presents in teens
-- Clinically significant? Yes

Because of these issues, stromal cysts require treatment in most cases!
Pediatric Iris Cysts

Pupillary cysts
--Pigment epi cysts at the...pupil border
--Presents in...infancy

Iris pigment epithelium cysts
--2° to separation of...epithelial layers
--Presents in...teens
--Clinically significant? No

Iris pigment epithelium cysts
--Due to stromal sequestration of...epithelial tissue
during embryogenesis
--Presents in...infancy--Cysts contain...goblet cells, may cause enlargement
--Result in a number of problems:
---Obstruct...visual axis
---Secondary...glaucoma
---Induce...corneal decompensation
---Cyst leakage → chronic iritis

How are stromal cysts treated?

Because of these issues, stromal cysts require treatment in most cases!

Stromal cysts
--Cysts contain...goblet cells, may cause enlargement

Other cysts
---Obstruct...visual axis
---Secondary...glaucoma
---Induce...corneal decompensation
---Cyst leakage → chronic iritis
Pediatric Iris Cysts

How are stromal cysts treated?
There are multiple approaches; each has its advantages and disadvantages. **Photodisruption** of the cyst is simple and noninvasive, but risks inciting iritis and/or glaucoma when the pro-inflammatory contents of the cyst are released into the AC. Because of this, the Peds book recommends **surgical excision**. Other sources recommend the less-invasive **aspiration with absolute alcohol injection**.

**Iris pigment epithelium cysts**
- 2° to separation of epithelial layers
- Presents in...teens
- Clinically significant? No

**Pupillary cysts**
- Pigment epi cysts at the...pupil border
- Presents in...infancy

Because of these issues, stromal cysts require treatment in most cases!

- Cysts contain...goblet cells, may cause enlargement
- Obstruct...visual axis
- Secondary...glaucoma
- Induce...corneal decompensation
- Cyst leakage → chronic iritis
Pediatric Iris Cysts

Primary

Epithelial

Stromal

Secondary

Pediatric Iris Cysts

Stromal cysts

--Due to stromal sequestration of epithelial tissue during embryogenesis
--Presents in infancy
--Cysts contain goblet cells, may cause enlargement
--Result in a number of problems:
  --Obstruct visual axis
  --Secondary glaucoma
  --Induce corneal decompensation
  --Cyst leakage → chronic iritis

Which is more common—epithelial, or stromal cysts?

Epithelial, by a mile
Pediatric Iris Cysts

Primary

- Epithelial
- Stromal

Which is more common—epithelial, or stromal cysts? Epithelial, by a mile

Secondary

- Iris pigment epithelium cysts
  - Pigment epi cysts at the pupil border
  - Presents in infancy

Iris pigment epithelium cysts
  - Due to separation of epithelial layers
  - Presents in teens
  - Clinically significant? No

Stromal cysts
  - Due to stromal sequestration of epithelial tissue during embryogenesis
  - Presents in infancy
  - Cysts contain goblet cells, may cause enlargement
  - Result in a number of problems:
    - Obstruct visual axis
    - Secondary glaucoma
    - Induce corneal decompensation
    - Cyst leakage → chronic iritis
Pediatric Iris Cysts

**Primary Iris Cysts**

- **Stromal cysts**
  - Due to stromal sequestration of epithelial tissue during embryogenesis
  - Presents in infancy
  - Cysts contain goblet cells, may cause enlargement
  - Result in a number of problems:
    - Obstruct visual axis
    - Secondary glaucoma
    - Induce corneal decompensation
    - Cyst leakage → chronic iritis

- **Epithelial cysts**
  - Iris pigment epithelium cysts
    - 2° to separation of epithelial layers
    - Presents in teens
    - Clinically significant? No

**Secondary Iris Cysts**

- Usually due to [drug]
Pediatric Iris Cysts

Primary

Epithelial

Stromal

Secondary

Secondary cysts
--Usually due to… echothiophate

Iris pigment epithelium cysts
--Due to stromal sequestration of… epithelial tissue during embryogenesis
--Presents in… infancy
--Cysts contain… goblet cells, may cause enlargement
--Result in a number of problems:
  --Obstruct… visual axis
  --Secondary… glaucoma
  --Induce… corneal decompensation
  --Cyst leakage → chronic iritis

Pupillary cysts
--Pigment epi cysts at the… pupil border
--Presents in… infancy

Iris pigment epithelium cysts
--2o to separation of… epithelial layers
--Presents in… teens
--Clinically significant? No

Pupillary cysts
--Pigment epi cysts at the… pupil border
--Presents in… infancy
Pediatric Iris Cysts

Primary Iris Cysts
- Epithelial
  - Pupillary cysts
  - Iris pigment epithelium cysts
- Stromal

Secondary Iris Cysts
- Usually due to...echothiophate

What is the trade name for echothiophate? Phospholine

What sort of drug is it? A parasympathomimetic
What is its mechanism of action? It is an acetylcholinesterase inhibitor

Ocular condition echothiophate is used to treat? Accommodative ET
Pediatric Iris Cysts

Primary

Epithelial

Pupillary cysts

Iris pigment epithelium cysts

Stromal

Secondary

Secondary cysts

--Usually due to... echothiophate

What is the trade name for echothiophate?
Phospholine

Secondary cysts

--Usually due to... echothiophate

What is the trade name for echothiophate?
Phospholine

Pediatric Iris Cysts

Epithelial

Iris pigment epithelium cysts

Pupillary cysts

--Pigment epi cysts at the... pupil border
--Presents in... infancy

Iris pigment epithelium cysts

--2o to separation of... epithelial layers
--Presents in... teens
--Clinically significant? No
Pediatric Iris Cysts

Primary Iris Cysts

Epithelial

Iris pigment epithelium cysts

Pupillary cysts -- Pigment epi cysts at the pupil border -- Presents in infancy

Stromal

Secondary Iris Cysts

Secondary cysts -- Usually due to echothiophate

What is the trade name for echothiophate?
Phospholine

What sort of drug is it?
A parasympathomimetic

What is its mechanism of action?
It is an acetylcholinesterase inhibitor

What ocular condition is echothiophate used to treat?
Accommodative ET
Pediatric Iris Cysts

Primary Iris Cysts

- Epithelial
  - Pupillary cysts
    - Pigment epithelial cysts
      - 2° to separation of epithelial layers
        - Presents in teens
        - Clinically significant? No
- Stromal
  - Iris pigment epithelium cysts

Secondary Iris Cysts

- Usually due to echothiophate

Echothiophate
- What is the trade name? Phospholine
- What sort of drug is it? A parasympathomimetic
- What is the mechanism of action? Acetylcholinesterase inhibitor
- Used to treat Accommodative ET
Pediatric Iris Cysts

Primary Iris Cysts

Epithelial
- Pupillary cysts
  -- Pigment epithelium cysts
    -- Secondary cysts
      -- Usually due to echothiophate
- Iris pigment epithelium cysts
  -- 2° to separation of epithelial layers

Stromal
- Primary

Secondary Iris Cysts

Secondary cysts
- Usually due to echothiophate

Pediatric Iris Cysts

What is the trade name for echothiophate?
Phospholine

What sort of drug is it?
A parasympathomimetic

What is its mechanism of action?
It is an acetylcholinesterase inhibitor

What ocular condition is echothiophate used to treat?
Accommodative ET
Pediatric Iris Cysts

**Primary Iris Cysts**

- **Epithelial**
  - **Pupillary cysts**
    - Pigment epi cysts at the pupil border
    - Presents in infancy
  - **Iris pigment epithelium cysts**
    - 2o to separation of epithelial layers
    - Presents in teens
    - Clinically significant? No

- **Stromal Iris Cysts**

**Secondary Iris Cysts**

- Usually due to echothiophate

**Pediatric Iris Cysts**

**Secondary cysts**

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

- Phospholine

**What sort of drug is it?**

- A parasympathomimetic

**What is its mechanism of action?**

- It is an acetylcholinesterase inhibitor
Pediatric Iris Cysts

Primary

Epithelial

Pupillary cysts

Iris pigment epithelium cysts

--Pigment epi cysts at the...pupil border
--Presents in...infancy

Secondary

Stromal

Secondary cysts

--Usually due to...echothiophate

What is the trade name for echothiophate?

Phospholine

What sort of drug is it?

A parasympathomimetic

What is its mechanism of action?

It is an acetylcholinesterase inhibitor

What ocular condition is echothiophate used to treat?
Pediatric Iris Cysts

Primary

Epithelial

Iris pigment epithelium cysts

Pupillary cysts

--Pigment epi cysts at the pupil border
--Presents in infancy

Stromal

Secondary

Secondary cysts
--Usually due to echothiophate

What is the trade name for echothiophate?
Phospholine

What sort of drug is it?
A parasympathomimetic

What is its mechanism of action?
It is an acetylcholinesterase inhibitor

What ocular condition is echothiophate used to treat?
Accommodative ET
Pediatric Iris Cysts

Primary

Epithelial

Pupillary cysts
--Pigment epi cysts at the…pupil border
--Presents in…infancy

Iris pigment epithelium cysts
--2° to separation of…epithelial layers
--Presents in…teens
--Clinically significant? No

Secondary

Stromal

Secondary cysts
--Usually due to…echothiophate
--Can prophylax with…[drug]

Stromal cysts
--Due to stromal sequestration of…epithelial tissue during embryogenesis
--Presents in…infancy
--Cysts contain…goblet cells, may cause enlargement
--Result in a number of problems:
  --Obstruct…visual axis
  --Secondary…glaucoma
  --Induce…corneal decompensation
  --Cyst leakage → chronic iritis
Pediatric Iris Cysts

Primary

Epithelial

Pupillary cysts

Iris pigment epithelium cysts

Secondary

Stromal

Secondary cysts
--Usually due to...echothiophate
--Can prophylax with...phenylephrine

Stromal cysts
--Due to stromal sequestration of...epithelial tissue during embryogenesis
--Presents in...infancy
--Cysts contain...goblet cells, may cause enlargement
--Result in a number of problems:
  --Obstruct...visual axis
  --Secondary...glaucoma
  --Induce...corneal decompensation
  --Cyst leakage → chronic iritis

Pupillary cysts
--Pigment epi cysts at the...pupil border
--Presents in...infancy

Iris pigment epithelium cysts
--2° to separation of...epithelial layers
--Presents in...teens
--Clinically significant? No
Name 3 types of iris nodules found in the pediatric population

Mnemonic forthcoming…
Name 3 types of iris nodules found in the pediatric population

- L
- B
- J

Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules…
Name 3 types of iris nodules found in the pediatric population

- **L**isch nodules
- **B**rushfield spots
- **J**XG nodules

Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules…
Name 3 types of iris nodules found in the pediatric population:
- **Lisch nodules**
- **Brushfield spots**
- **JXG nodules**

Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules…

Lisch nodules are most strongly associated with what congenital condition?

- **NF1**

Is it associated with NF2?
- Yes, but the relationship is far weaker—Lisch nodules occur in NF2, but so sporadically that they are not expected

What is the prevalence of Lisch nodules in NF1?
- The rule-of-thumb is that Lisch nodule prevalence equals the age of the patient times 10. Thus, 50% of 5 year olds will have them, 60% of 6 year olds, etc. At age 10 and beyond, essentially 100% of NF1 patients have Lisch nodules.

Are Lisch nodules clinically significant?
- No; their only significance is as a diagnostic marker for NF1
Name 3 types of iris nodules found in the pediatric population:

- **Lisch nodules**
- **Brushfield spots**
- **JXG nodules**

Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules…

Lisch nodules are most strongly associated with what congenital condition?
Neurofibromatosis type 1 (NF1)

Are Lisch nodules clinically significant?
No; their only significance is as a diagnostic marker for NF1

Lisch nodules are most strongly associated with what congenital condition?
Neurofibromatosis type 1 (NF1)
Name 3 types of iris nodules found in the pediatric population:

- Lisch nodules
- Brushfield spots
- JXG nodules

Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules...

Lisch nodules are most strongly associated with what congenital condition?

Neurofibromatosis type 1 (NF1)

What is the eponymous name for NF1?

von Recklinghausen's disease

In a word, what sort of condition is NF1?

A phakomatosis
Name 3 types of iris nodules found in the pediatric population:

- Lisch nodules
- Brushfield spots
- JXG nodules

Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules...

Lisch nodules are most strongly associated with what congenital condition?

**Neurofibromatosis type 1 (NF1)**

What is the eponymous name for NF1? von Recklinghausen’s disease

Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules...
Name 3 types of iris nodules found in the pediatric population:
- Lisch nodules
- Brushfield spots
- JXG nodules

Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules...

Lisch nodules are most strongly associated with what congenital condition?

**Neurofibromatosis type 1 (NF1)**

What is the eponymous name for NF1?
von Recklinghausen’s disease

In a word, what sort of condition is NF1?

A phakomatosis
Lisch nodules are most strongly associated with what congenital condition? 
**Neurofibromatosis type 1 (NF1)**

What is the eponymous name for NF1? von Recklinghausen’s disease

In a word, what sort of condition is NF1? A phakomatosis

Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules...
Name 3 types of iris nodules found in the pediatric population:

- **Lisch nodules**
- **Brushfield spots**
- **JXG nodules**

**Lisch nodules** are most strongly associated with what congenital condition?

**Neurofibromatosis type 1 (NF1)**

What is the eponymous name for NF1?

von Recklinghausen’s disease

In a word, what sort of condition is NF1?

A phakomatosis

Briefly, what is a phakomatosis?

Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules…
Lisch nodules are most strongly associated with what congenital condition?  
**Neurofibromatosis type 1 (NF1)**

What is the eponymous name for NF1?  
von Recklinghausen’s disease

In a word, what sort of condition is NF1?  
**A** phakomatosis

**Briefly, what is a phakomatosis?**  
A congenital condition involving hamartomatous lesions of multiple organ systems, usually including the **CNS**, **eyes** and **skin**

Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules…
Name 3 types of iris nodules found in the pediatric population:

- Lisch nodules
- Brushfield spots
- JXG nodules

Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules...

Lisch nodules are most strongly associated with what congenital condition?

**Neurofibromatosis type 1 (NF1)**

What is the eponymous name for NF1?

von Recklinghausen’s disease

In a word, what sort of condition is NF1?

A phakomatosis

Briefly, what is a phakomatosis?

A congenital condition involving hamartomatous lesions of multiple organ systems, usually including the CNS, eyes, and skin.

What is the prevalence of Lisch nodules in NF1?

The rule-of-thumb is that Lisch nodule prevalence equals the age of the patient times 10. Thus, 50% of 5 year olds will have them, 60% of 6 year olds, etc. At age 10 and beyond, essentially 100% of NF1 patients have Lisch nodules.

Are Lisch nodules clinically significant?

No; their only significance is as a diagnostic marker for NF1.
Name 3 types of iris nodules found in the pediatric population:

- Lisch nodules
- Brusfield spots
- JXG nodules

Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules...

**Lisch nodules are most strongly associated with what congenital condition?**

**Neurofibromatosis type 1 (NF1)**

**What is the eponymous name for NF1?**

von Recklinghausen’s disease

**In a word, what sort of condition is NF1?**

A **phakomatosis**

**Briefly, what is a phakomatosis?**

A congenital condition involving hamartomatous lesions of multiple organ systems, usually including the CNS, eyes and skin

**By what more-descriptive name does the BCSC Peds book refer to them?**
Name 3 types of iris nodules found in the pediatric population:

- Lisch nodules
- Brushfield spots
- JXG nodules

Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules…

Lisch nodules are most strongly associated with what congenital condition?

**Neurofibromatosis type 1 (NF1)**

What is the eponymous name for NF1? von Recklinghausen’s disease

In a word, what sort of condition is NF1?

A **phakomatosis**

Briefly, what is a phakomatosis?

A congenital condition involving hamartomatous lesions of multiple organ systems, usually including the CNS, eyes, and skin

By what more-descriptive name does the BCSC Peds book refer to them?

As neuro-

Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules…
Name 3 types of iris nodules found in the pediatric population:

- Lisch nodules
- Brushfield spots
- JXG nodules

Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules...

**Lisch nodules are most strongly associated with what congenital condition?**

**Neurofibromatosis type 1 (NF1)**

**What is the eponymous name for NF1?**

von Recklinghausen’s disease

**In a word, what sort of condition is NF1?**

A **phakomatosis**

**Briefly, what is a phakomatosis?**

A congenital condition involving hamartomatous lesions of multiple organ systems, usually including the **CNS, eyes** and skin

**By what more-descriptive name does the BCSC Peds book refer to them?**

As **neuro-oculo**
Name 3 types of iris nodules found in the pediatric population:

- **Lisch nodules**
- **B Rushfield spots**
- **JXG nodules**

Think of President Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules…

*Lisch nodules are most strongly associated with what congenital condition?*
- **Neurofibromatosis type 1 (NF1)**

What is the eponymous name for NF1?
- von Recklinghausen’s disease

In a word, what sort of condition is NF1?
- A **phakomatosis**

_Briefly, what is a phakomatosis?_  
A congenital condition involving hamartomatous lesions of multiple organ systems, usually including the **CNS, eyes and skin**

By what more-descriptive name does the BCSC Peds book refer to them?  
As **neuro-oculocutaneous** syndromes
Name 3 types of iris nodules found in the pediatric population:

- **Lisch nodules**
- **Brushfield spots**
- **JXG nodules**

Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules…

*Lisch nodules are most strongly associated with what congenital condition? Neurofibromatosis type 1 (NF1)*

*Is it associated with NF2?*

No; their only significance is as a diagnostic marker for NF1.
**A**

- Name 3 types of iris nodules found in the pediatric population:
  - **Lisch nodules**
  - **Brushfield spots**
  - **JXG nodules**

*Lisch nodules are most strongly associated with what congenital condition?*
Neurofibromatosis type 1 (NF1)

*Is it associated with NF2?*
Yes, but the relationship is far weaker—Lisch nodules occur in NF2, but so sporadically that they are not expected.

*Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules…*
Name 3 types of iris nodules found in the pediatric population:
- **Lisch nodules**
- Brushfield spots
- **JXG nodules**

Lisch nodules are most strongly associated with what congenital condition?
- Neurofibromatosis type 1 (NF1)

Is it associated with NF2?
- Yes, but the relationship is far weaker—Lisch nodules occur in NF2, but so sporadically that they are not expected

What is the prevalence of Lisch nodules in NF1?
- The rule-of-thumb is that Lisch nodule prevalence equals the age of the patient times 10. Thus, 50% of 5 year olds will have them, 60% of 6 year olds, etc. At age 10 and beyond, essentially 100% of NF1 patients have Lisch nodules.

Are Lisch nodules clinically significant?
- No; their only significance is as a diagnostic marker for NF1

Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules…
Lisch nodules are most strongly associated with what congenital condition? Neurofibromatosis type 1 (NF1)

Is it associated with NF2?
Yes, but the relationship is far weaker—Lisch nodules occur in NF2, but so sporadically that they are not expected

What is the prevalence of Lisch nodules in NF1?
The rule-of-thumb is that Lisch nodule prevalence equals the age of the patient times 10. Thus, 50% of 5 year olds will have them, 60% of 6 year olds, etc. At age 10 and beyond, essentially 100% of NF1 patients have Lisch nodules.
Name 3 types of iris nodules found in the pediatric population:

- **Lisch nodules**
- **Brushfield spots**
- **JXG nodules**

**Lisch nodules are most strongly associated with what congenital condition?**

Neurofibromatosis type 1 (NF1)

**Is it associated with NF2?**

Yes, but the relationship is far weaker—Lisch nodules occur in NF2, but so sporadically that they are not expected.

**What is the prevalence of Lisch nodules in NF1?**

The rule-of-thumb is that Lisch nodule prevalence equals the age of the patient times 10. Thus, 50% of 5 year olds will have them, 60% of 6 year olds, etc. At age 10 and beyond, essentially 100% of NF1 patients have Lisch nodules.

**Are Lisch nodules clinically significant?**

No; their only significance is as a diagnostic marker for NF1.

Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules…
Name 3 types of iris nodules found in the pediatric population:

- **Lisch nodules**
- Brushfield spots
- JXG nodules

**Lisch nodules are most strongly associated with what congenital condition?**
Neurofibromatosis type 1 (NF1)

**Is it associated with NF2?**
Yes, but the relationship is far weaker—Lisch nodules occur in NF2, but so sporadically that they are not expected.

**What is the prevalence of Lisch nodules in NF1?**
The rule-of-thumb is that Lisch nodule prevalence equals the age of the patient times 10. Thus, 50% of 5 year olds will have them, 60% of 6 year olds, etc. At age 10 and beyond, essentially 100% of NF1 patients have Lisch nodules.

**Are Lisch nodules clinically significant?**
No; their only significance is as a diagnostic marker for NF1.

Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules…
Name 3 types of iris nodules found in the pediatric population

- Lisch nodules
- Brushfield spots
- JXG nodules
- Iris

Another sort of iris nodule needs mentioning here, because it is in the DDx for Lisch nodules. What is it?

Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules…
Name 3 types of iris nodules found in the pediatric population

- *Lisch nodules*
- *Brushfield spots*
- *JXG nodules*
- *Iris mammillations*

Another sort of iris nodule needs mentioning here, because it is in the DDx for Lisch nodules. What is it?

*Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules…*
Name 3 types of iris nodules found in the pediatric population:

- **Lisch nodules**
- **Brushfield spots**
- **JXG nodules**
- **Iris mammillations**

Another sort of iris nodule needs mentioning here, because it is in the DDx for Lisch nodules. What is it?

Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules…
Name 3 types of iris nodules found in the pediatric population:

- **Lisch nodules**
- **Brushfield spots**
- **JXG nodules**

Another sort of iris nodule needs mentioning here, because it is in the DDx for Lisch nodules. What is it?

Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules…

Mammillations? Aren’t those a CNS thingamajig? You’re thinking of the _two words_, paired structures that are part of the limbic system.
Name 3 types of iris nodules found in the pediatric population:

- **Lisch nodules**
- **Brushfield spots**
- **JXG nodules**
- **Iris mammillations**

Another sort of iris nodule needs mentioning here, because it is in the DDx for Lisch nodules. What is it?

Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules…
Name 3 types of iris nodules found in the pediatric population:

- **Lisch nodules**
- **Brushfield spots**
- **JXG nodules**
- **Iris mammillations**

Another sort of iris nodule needs mentioning here, because it is in the DDx for Lisch nodules. What is it?

Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules...
Name 3 types of iris nodules found in the pediatric population:

- **Lisch nodules**
- **Brushfield spots**
- **JXG nodules**
- **Iris mammillations**

Another sort of iris nodule needs mentioning here, because it is in the DDx for Lisch nodules. What is it?

_Mammillations? Aren’t those a CNS thingamajig?_ You’re thinking of the _mammillary bodies_, paired structures that are part of the limbic system.

_OK, then what are iris mammillations?_ Tiny pigmented nodules which, when present, are found in vast numbers diffusely scattered across the iris surface.

*Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules...*
- Name 3 types of iris nodules found in the pediatric population:
  - **Lisch nodules**
  - **Brushfield spots**
  - **JXG nodules**
  - **Iris mammillations**

  *Mammillations? Aren’t those a CNS thingamajig? You’re thinking of the mammillary bodies, paired structures that are part of the limbic system*

  *OK, then what are iris mammillations? Tiny pigmented nodules which, when present, are found in vast numbers diffusely scattered across the iris surface*

  *Are they unilateral, or bilateral?*

  *Another sort of iris nodule needs mentioning here, because it is in the DDx for Lisch nodules. What is it?*

  *Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules...*
Name 3 types of iris nodules found in the pediatric population:

- **Lisch nodules**
- **Brushfield spots**
- **JXG nodules**
- **Iris mammillations**

Another sort of iris nodule needs mentioning here, because it is in the DDx for Lisch nodules. What is it?

Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules…
Name 3 types of iris nodules found in the pediatric population:

- **Lisch nodules**
- **Brushfield spots**
- **JXG nodules**
- **Iris mammillations**

Mammillations? Aren’t those a CNS thingamajig? You’re thinking of the *mammillary bodies*, paired structures that are part of the limbic system.

OK, then *what are iris mammillations*? Tiny pigmented nodules which, when present, are found in vast numbers diffusely scattered across the iris surface.

Are they unilateral, or bilateral? Usually unilateral, but bilaterality occurs frequently enough that it can’t be used to rule them out.

Are they associated with NF1?

Another sort of *iris nodule* needs mentioning here, because it is in the DDx for Lisch nodules. What is it?

Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having *iris nodules*…
Name 3 types of iris nodules found in the pediatric population:

- **Lisch nodules**
- Brushfield spots
- **JXG nodules**
- **Iris mammillations**

Another sort of iris nodule needs mentioning here, because it is in the DDx for Lisch nodules. What is it?

Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules
Name 3 types of iris nodules found in the pediatric population:

- **Lisch nodules**
- **Brushfield spots**
- **JXG nodules**
- **Iris mammillations**

Another sort of iris nodule needs mentioning here, because it is in the DDx for Lisch nodules. What is it?

Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules…
Name 3 types of iris nodules found in the pediatric population:

- **Lisch nodules**
- **Brushfield spots**
- **JXG nodules**
- **Iris mammillations**

Another sort of iris nodule needs mentioning here, because it is in the DDx for Lisch nodules. **What is it?**

Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules...
Name 3 types of iris nodules found in the pediatric population:

- **Lisch nodules**
- **Brushfield spots**
- **JXG nodules**

**Iris mammillations**

Another sort of iris nodule needs mentioning here, because it is in the DDx for Lisch nodules. What is it?

*Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules...*
Name 3 types of iris nodules found in the pediatric population:

- **Lisch nodules**
- **Brushfield spots**
- **JXG nodules**
- **Iris mammillations**

Another sort of iris nodule needs mentioning here, because it is in the DDx for Lisch nodules. What is it?

Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules…
Name 3 types of iris nodules found in the pediatric population:

- Lisch nodules
- Brushfield spots
- JXG nodules
- Iris mammillations

Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules…

Mammillations? Aren't those a CNS thingamajig? You're thinking of the mammillary bodies, paired structures that are part of the limbic system.

OK, then what are iris mammillations? Tiny pigmented nodules which, when present, are found in vast numbers diffusely scattered across the iris surface.

Are they usually unilateral? Yes, but they occur bilaterally frequently enough that it can't be used to rule them out.

Are they associated with NF1? Yes (albeit not nearly as strongly as Lisch nodules).

In addition to NF1, iris mammillations have another important association. What is it? Oculodermal melanocytosis, aka nevus of Ota.

What is oculodermal melanocytosis/nevus of Ota? Ocular melanocytosis + hyperpigmentation of the ipsilateral periorbital skin.

What racial group is most likely to be affected? Asians.

Another sort of iris nodule needs mentioning here, because it is in the DDx for Lisch nodules. What is it?
Name 3 types of iris nodules found in the pediatric population:

- Lisch nodules
- Rushfield spots
- JXG nodules

Are they unilateral, or bilateral? Usually unilateral, but bilaterality occurs frequently enough that it can't be used to rule them out.

Are they associated with NF1? Yes (albeit not nearly as strongly as Lisch nodules).

Another sort of iris nodule needs mentioning here, because it is in the DDx for Lisch nodules. What is it?

What is oculodermal melanocytosis/nevus of Ota? Ocular melanocytosis + hyperpigmentation of the ipsilateral periorbital skin. Are they associated with NF1 and oculodermal melanocytosis? Yes.

In addition to NF1, iris mammillations have another important association. What is it? It is oculodermal melanocytosis, aka nevus of Ota.

Another sort of iris nodule needs mentioning here, because it is in the DDx for Lisch nodules. What is it?

Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules…
Name 3 types of iris nodules found in the pediatric population:

- **Lisch nodules**
- **Buchwald spots**
- **JXG nodules**

Iris mammillations? Aren’t those a CNS thingamajig? You’re thinking of the **mammillary bodies**, paired structures that are part of the limbic system.

OK, then what are iris mammillations? Tiny pigmented nodules which, when present, are found in vast numbers diffusely scattered across the iris surface.

Are they usually unilateral? Usually, though this isn’t always the case.

Are they associated with NF1? They are, albeit not nearly as strongly as Lisch nodules.

Another sort of iris nodule needs mentioning here, because it is in the DDx for Lisch nodules. What is it?

Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules…
Name 3 types of iris nodules found in the pediatric population:

- Lisch nodules
- B Russell spots
- JXG nodules
- Iris mammillations

Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules…

Mammillations? Aren’t those a CNS thingamajig? You’re thinking of the mammillary bodies, paired structures that are part of the limbic system.

OK, then what are iris mammillations? Tiny pigmented nodules which, when present, are found in vast numbers diffusely scattered across the iris surface.

Are they usually unilateral or bilateral? Usually unilateral, but bilaterality occurs frequently enough that it can’t be used to rule them out.

Are they associated with NF1? Yes (albeit not nearly as strongly as Lisch nodules).

Another sort of iris nodule needs mentioning here, because it is in the DDx for Lisch nodules. What is it?

Oculodermal melanocytosis, aka nevus of Ota: Ocular melanocytosis + hyperpigmentation of the ipsilateral periorbital skin.

What racial group is most likely to be affected? Asians.

In addition to NF1, iris mammillations have another important association. What is it?

Oculodermal melanocytosis, aka oculodermal melanocytosis.

What is oculodermal melanocytosis/nevus of Ota? Ocular melanocytosis + hyperpigmentation of the ipsilateral periorbital skin.

What racial group is most likely to be affected? Asians.

Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules…
Name 3 types of iris nodules found in the pediatric population:

- Lisch nodules
- Rushfield spots
- JXG nodules

Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules…

Mammillations? Aren't those a CNS thingamajig? You're thinking of the mammillary bodies, paired structures that are part of the limbic system.

OK, then what are iris mammillations?

Tiny pigmented nodules which, when present, are found in vast numbers diffusely scattered across the iris surface.

Are they unilateral, or bilateral?

Usually unilateral, but bilaterality occurs frequently enough that it can't be used to rule them out.

Are they associated with NF1?

Yes (albeit not nearly as strongly as Lisch nodules).

Another sort of iris nodule needs mentioning here, because it is in the DDx for Lisch nodules. What is it?

In addition to NF1, iris mammillations have another important association. What is it?

Oculodermal melanocytosis, aka nevus of Ota and ocular dermal melanocytosis.

What is oculodermal melanocytosis/nevus of Ota?

Ocular melanocytosis + hyperpigmentation of the ipsilateral periorbital skin.

What racial group is most likely to be affected?

Asians.

What is ocular melanocytosis?

A pigmented lesion of the episclera.

At what age does it present?

It is congenital, so birth.

Who get it?

People of pigmented ethnicity, i.e., blacks, Hispanics, Asians.

Is it unilateral, or bilateral?

Most cases are unilateral.

Is it associated with ocular morbidity?

Yes, 10% of cases develop ipsilateral glaucoma.

What percent of ocular melanocytosis cases have oculo-dermal melanocytosis?

About half.

Does ocular melanocytosis possess malignant potential?

Only in whites.
Name 3 types of iris nodules found in the pediatric population:

- Lisch nodules
- Budgefield spots
- JXG nodules

Iris mammillations? Aren’t those a CNS thingamajig? You’re thinking of the mammillary bodies, paired structures that are part of the limbic system. OK, then what are iris mammillations? Tiny pigmented nodules which, when present, are found in vast numbers diffusely scattered across the iris surface.

Are they unilateral, or bilateral? Usually unilateral, but bilaterality occurs frequently enough that it can’t be used to rule them out.

Are they associated with NF1? Yes (albeit not nearly as strongly as Lisch nodules).

Another sort of iris nodule needs mentioning here, because it is in the DDx for Lisch nodules. What is it? In addition to NF1, iris mammillations have another important association. What is it? Oculodermal melanocytosis, aka nevus of Ota.

What is oculodermal melanocytosis/nevus of Ota? Ocular melanocytosis + hyperpigmentation of the ipsilateral periorbital skin.

What racial group is most likely to be affected? Asians.

What is ocular melanocytosis? A pigmented lesion of the episclera. At what age does it present? It is congenital, so birth. Who get it? People of pigmented ethnicity, ie, blacks, Hispanics, Asians. Is it unilateral, or bilateral? Most cases are unilateral. Is it associated with ocular morbidity? Yes, 10% of cases develop ipsilateral glaucoma. What percent of ocular melanocytosis cases have oculo-dermal melanocytosis? About half. Does ocular melanocytosis possess malignant potential? Only in whites.
Name 3 types of iris nodules found in the pediatric population:

- Lisch nodules
- Berson spots
- JXG nodules

Iris mammillations? Aren't those a CNS thingamajig?

You're thinking of the mammillary bodies, paired structures that are part of the limbic system.

OK, then what are iris mammillations?

Tiny pigmented nodules which, when present, are found in vast numbers diffusely scattered across the iris surface.

Are they unilateral, or bilateral?

Usually unilateral, but bilaterality occurs frequently enough that it can't be used to rule them out.

Are they associated with NF1?

Yes (albeit not nearly as strongly as Lisch nodules).

Another sort of iris nodule needs mentioning here, because it is in the DDx for Lisch nodules. What is it?

In addition to NF1, iris mammillations have another important association. What is it?

Oculodermal melanocytosis, aka nevus of Ota and ocular melanocytosis.

What is oculodermal melanocytosis/nevus of Ota?

Ocular melanocytosis + hyperpigmentation of the ipsilateral periorbital skin.

What racial group is most likely to be affected?

Asians.

What is ocular melanocytosis?

A pigmented lesion of the episclera.

At what age does it present?

It is congenital, so birth.

Who get it?

People of pigmented ethnicity, ie, blacks, Hispanics, Asians.

Is it unilateral, or bilateral?

Most cases are unilateral.

Is it associated with ocular morbidity?

Yes, 10% of cases develop ipsilateral glaucoma.

What percent of ocular melanocytosis cases have oculo-dermal melanocytosis?

About half.

Does ocular melanocytosis possess malignant potential?

Only in whites.
Name 3 types of iris nodules found in the pediatric population:

- Lisch nodules
- Buschfield spots
- JXG nodules

Mammillations? Aren’t those a CNS thingamajig? You’re thinking of the mammillary bodies, paired structures that are part of the limbic system.

What are iris mammillations?

Tiny pigmented nodules which, when present, are found in vast numbers diffusely scattered across the iris surface.

Are they unilateral, or bilateral?

Usually unilateral, but bilaterality occurs frequently enough that it can’t be used to rule them out.

Are they associated with NF1?

Yes (albeit not nearly as strongly as Lisch nodules).

Another sort of iris nodule needs mentioning here, because it is in the DDx for Lisch nodules. What is it?

In addition to NF1, iris mammillations have another important association. What is it?

Oculodermal melanocytosis, aka nevus of Ota and… oculodermal melanocytosis.

What is oculodermal melanocytosis/nevus of Ota?

Ocular melanocytosis + hyperpigmentation of the ipsilateral periorbital skin.

What racial group is most likely to be affected?

Asians.

What is ocular melanocytosis?

A pigmented lesion of the episclera.

At what age does it present?

It is congenital, so birth.

Who get it?

People of pigmented ethnicity, ie, blacks, Hispanics, Asians.

Is it unilateral, or bilateral?

Most cases are unilateral.

Is it associated with ocular morbidity?

Yes, 10% of cases develop ipsilateral glaucoma.

What percent of ocular melanocytosis cases have oculo-dermal melanocytosis?

About half.

Does ocular melanocytosis possess malignant potential?

Only in whites.
Name 3 types of iris nodules found in the pediatric population:

- Lisch nodules
- Rushfield spots
- JXG nodules

Iris mammillations? Aren't those a CNS thingamajig? You're thinking of the mammillary bodies, paired structures that are part of the limbic system.

OK, then what are iris mammillations? Tiny pigmented nodules which, when present, are found in vast numbers diffusely scattered across the iris surface. Are they unilateral, or bilateral? Usually unilateral, but bilaterality occurs frequently enough that it can't be used to rule them out. Are they associated with NF1? Yes (albeit not nearly as strongly as Lisch nodules).

Another sort of iris nodule needs mentioning here, because it is in the DDx for Lisch nodules. What is it? In addition to NF1, iris mammillations have another important association. What is it? Oculodermal melanocytosis, aka nevus of Ota and... oculodermal melanocytosis.

What is oculodermal melanocytosis/nevus of Ota? Ocular melanocytosis + hyperpigmentation of the ipsilateral periorbital skin. What racial group is most likely to be affected? Asians.

What is ocular melanocytosis? A pigmented lesion of the episclera. At what age does it present? It is congenital, so birth. Who get it? People of pigmented ethnicity, ie, blacks, Hispanics, Asians. Is it unilateral, or bilateral? Most cases are unilateral. Is it associated with ocular morbidity? Yes, 10% of cases develop ipsilateral glaucoma. What percent of ocular melanocytosis cases have oculo-dermal melanocytosis? About half. Does ocular melanocytosis possess malignant potential? Only in whites.
Name 3 types of iris nodules found in the pediatric population:

- Lisch nodules
- Rushfield spots
- JXG nodules

Iris mammillations: These are tiny pigmented nodules which, when present, are found in vast numbers diffusely scattered across the iris surface. They are usually unilateral, but bilaterality occurs frequently enough that it can't be used to rule them out.

Are they associated with NF1?

Yes (albeit not nearly as strongly as Lisch nodules)

Another sort of iris nodule needs mentioning here, because it is in the DDx for Lisch nodules. What is it?

Oculodermal melanocytosis, aka nevus of Ota and oculodermal melanocytosis.

What is oculodermal melanocytosis?

Ocular melanocytosis + hyperpigmentation of the ipsilateral periorbital skin.

What racial group is most likely to be affected?

Asians

What is ocular melanocytosis?

A pigmented lesion of the episclera

At what age does it present?

It is congenital, so birth

Who gets it?

People of pigmented ethnicity, ie, blacks, Hispanics, Asians

Is it unilateral, or bilateral?

Most cases are unilateral

Is it associated with ocular morbidity?

Yes, 10% of cases develop ipsilateral glaucoma

What percent of ocular melanocytosis cases have oculodermal melanocytosis?

About half

Does ocular melanocytosis possess malignant potential?

Only in whites.

Mammillations? Aren't those a CNS thingamajig?

You're thinking of the mammillary bodies, paired structures that are part of the limbic system.

Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules…
Name 3 types of iris nodules found in the pediatric population:

- **Lisch nodules**
- **Buchwald spots**
- **JXG nodules**

Think of President Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules…

Mammillations? Aren’t those a CNS thingamajig? You’re thinking of the **mamillary bodies**, paired structures that are part of the limbic system.

OK, then what are iris mammillations? Tiny pigmented nodules which, when present, are found in vast numbers diffusely scattered across the iris surface.

Are they unilateral, or bilateral? Usually unilateral, but bilaterality occurs frequently enough that it can’t be used to rule them out.

Are they associated with NF1? Yes (albeit not nearly as strongly as Lisch nodules).

Another sort of iris nodule needs mentioning here, because it is in the DDx for Lisch nodules. What is it? In addition to NF1, iris mammillations have another important association. What is it? Oculodermal melanocytosis, aka nevus of Ota and… oculodermal melanocytosis.

What is oculodermal melanocytosis? Ocular melanocytosis + hyperpigmentation of the ipsilateral periorbital skin.

What racial group is most likely to be affected? Asians.

What is ocular melanocytosis? A pigmented lesion of the episclera.

At what age does it present? It is congenital, so birth.

Who gets it? People of pigmented ethnicity, ie, blacks, Hispanics, Asians.

Is it unilateral, or bilateral? Most cases are unilateral.

Is it associated with ocular morbidity? Yes, 10% of cases develop ipsilateral glaucoma.

What percent of ocular melanocytosis cases have oculo-dermal melanocytosis? About half.

Does ocular melanocytosis possess malignant potential? Only in whites.
Name 3 types of iris nodules found in the pediatric population:

- Lisch nodules
- Rushfield spots
- JXG nodules

Are mammillations and mammillary bodies the same thing?

Mammillations? Aren’t those a CNS thingamajig?

You’re thinking of the mammillary bodies, paired structures that are part of the limbic system.

OK, then what are iris mammillations?

Tiny pigmented nodules which, when present, are found in vast numbers diffusely scattered across the iris surface.

Are they unilateral, or bilateral?

Usually unilateral, but bilaterality occurs frequently enough that it can’t be used to rule them out.

Are they associated with NF1?

Yes (albeit not nearly as strongly as Lisch nodules).

Another sort of iris nodule needs mentioning here, because it is in the DDx for Lisch nodules. What is it?

In addition to NF1, iris mammillations have another important association. What is it?

Oculodermal melanocytosis, aka nevus of Ota and… oculodermal melanocytosis.

What is oculodermal melanocytosis/nevus of Ota?

Ocular melanocytosis + hyperpigmentation of the ipsilateral periorbital skin.

What racial group is most likely to be affected?

Asians.

What is ocular melanocytosis?

A pigmented lesion of the episclera.

At what age does it present?

It is congenital, so birth.

Who gets it?

People of pigmented ethnicity, ie, blacks, Hispanics, Asians.

Is it unilateral, or bilateral?

Most cases are unilateral.

Is it associated with ocular morbidity?

Yes, 10% of cases develop ipsilateral glaucoma.

What percent of ocular melanocytosis cases have oculo-dermal melanocytosis?

About half.

Does ocular melanocytosis possess malignant potential?

Only in whites.
Name 3 types of iris nodules found in the pediatric population:

- Lisch nodules
- Brushfield spots
- JXG nodules

Think of President Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules...

Mammillations? Aren't those a CNS thingamajig? You're thinking of the mammillary bodies, paired structures that are part of the limbic system.

OK, then what are iris mammillations?

Tiny pigmented nodules which, when present, are found in vast numbers diffusely scattered across the iris surface.

Are they unilateral, or bilateral?

Usually unilateral, but bilaterality occurs frequently enough that it can't be used to rule them out.

Are they associated with NF1?

Yes (albeit not nearly as strongly as Lisch nodules).

Another sort of iris nodule needs mentioning here, because it is in the DDx for Lisch nodules. What is it?

In addition to NF1, iris mammillations have another important association. What is it?

Oculodermal melanocytosis, aka nevus of Ota and oculodermal melanocytosis.

What is oculodermal melanocytosis/nevus of Ota?

Ocular melanocytosis + hyperpigmentation of the ipsilateral periorbital skin.

What racial group is most likely to be affected?

Asians.

What is ocular melanocytosis?

A pigmented lesion of the episclera.

At what age does it present?

It is congenital, so birth.

Who gets it?

People of pigmented ethnicity, ie, blacks, Hispanics, Asians.

Is it unilateral, or bilateral?

Most (~95%) cases are unilateral.

Is it associated with ocular morbidity?

Yes, 10% of cases develop ipsilateral glaucoma.

What percent of ocular melanocytosis cases have oculo dermal melanocytosis?

About half.

Does ocular melanocytosis possess malignant potential?

Only in whites.
Name 3 types of iris nodules found in the pediatric population:

- Lisch nodules
- Bashforth spots
- JXG nodules

Iris mammillations

Think of President Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules…

Mammillations? Aren’t those a CNS thingamajig? You’re thinking of the mammillary bodies, paired structures that are part of the limbic system.

OK, then what are iris mammillations?

Tiny pigmented nodules which, when present, are found in vast numbers diffusely scattered across the iris surface.

Are they unilateral, or bilateral?

Usually unilateral, but bilaterality occurs frequently enough that it can’t be used to rule them out.

Are they associated with NF1?

Yes (albeit not nearly as strongly as Lisch nodules).

Another sort of iris nodule needs mentioning here, because it is in the DDx for Lisch nodules. What is it?

In addition to NF1, iris mammillations have another important association. What is it?

Oculodermal melanocytosis, aka nevus of Ota and oculodermal melanocytosis.

What is oculodermal melanocytosis/nevus of Ota?

Ocular melanocytosis + hyperpigmentation of the ipsilateral periorbital skin.

What racial group is most likely to be affected?

Asians.

What is ocular melanocytosis?

A pigmented lesion of the episclera.

At what age does it present?

It is congenital, so birth.

Who gets it?

People of pigmented ethnicity, ie, blacks, Hispanics, Asians.

Is it unilateral, or bilateral?

Most (~95%) cases are unilateral.

Is it associated with ocular morbidity?

Yes, 10% of cases develop ipsilateral glaucoma.

What percent of ocular melanocytosis cases have oculo-dermal melanocytosis?

About half.

Does ocular melanocytosis possess malignant potential?

Only in whites.
Q/A

Name 3 types of iris nodules found in the pediatric population.

- Lisch nodules
- Brousset spots
- JXG nodules
- Iris mammillations

Mammillations? Aren't those a CNS thingamajig? You're thinking of the mammillary bodies, paired dorsoventrally across the diencephalon.

OK, then what are iris mammillations?

Tiny pigmented nodules which, when present, are found in vast numbers diffusely scattered across the iris surface.

Are they unilateral, or bilateral?

Usually unilateral, but bilaterality occurs frequently enough that it can't be used to rule them out.

Are they associated with NF1?

Yes (albeit not nearly as strongly as Lisch nodules).

Another sort of iris nodule needs mentioning here, because it is in the DDx for Lisch nodules. What is it?

Oculodermal melanocytosis, aka nevus of Ota. What is oculodermal melanocytosis/nevus of Ota?

Ocular melanocytosis + hyperpigmentation of the ipsilateral periorbital skin. What racial group is most likely to be affected?

Asians.

What is ocular melanocytosis?

A pigmented lesion of the episclera.

At what age does it present?

It is congenital, so birth.

Who gets it?

People of pigmented ethnicity, ie, blacks, Hispanics, Asians.

Is it unilateral, or bilateral?

Most (~95%) cases are unilateral.

Is it associated with ocular morbidity?

Yes, 10% of cases develop ipsilateral glaucoma.

What percent of ocular melanocytosis cases have oculo-dermal melanocytosis?

About half.

Does ocular melanocytosis possess malignant potential?

Only in whites.
Name 3 types of iris nodules found in the pediatric population:

- **Lisch nodules**
- **B Rushfield spots**
- **JXG nodules**
- **Iris mammillations**

Think of President Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules…

Mammillations? Aren’t those a CNS thingamajig? You’re thinking of the **mammillary bodies**, paired structures that are part of the limbic system. OK, then what are iris mammillations? Tiny pigmented nodules which, when present, are found in vast numbers diffusely scattered across the iris surface. Are they unilateral, or bilateral? Usually unilateral, but bilaterality occurs frequently enough that it can’t be used to rule them out. Are they associated with NF1? Yes (albeit not nearly as strongly as Lisch nodules).

Another sort of iris nodule needs mentioning here, because it is in the DDx for Lisch nodules. What is it? In addition to NF1, iris mammillations have another important association. What is it? Oculodermal melanocytosis, aka nevus of Ota. What is oculodermal melanocytosis/nevus of Ota? Ocular melanocytosis + hyperpigmentation of the ipsilateral periorbital skin. What racial group is most likely to be affected? Asians.

What is ocular melanocytosis? A pigmented lesion of the episclera. At what age does it present? It is congenital, so birth. Who gets it? People of pigmented ethnicity, ie, blacks, Hispanics, Asians. Is it unilateral, or bilateral? Most (~95%) cases are unilateral. Is it associated with ocular morbidity? Yes, 10% of cases develop ipsilateral glaucoma. Is it associated with other systemic conditions? Yes, it is often associated with oculodermal melanocytosis (nevus of Ota). What percent of ocular melanocytosis cases have oculo‐dermal melanocytosis? About half. Does ocular melanocytosis possess malignant potential? Only in whites.
Name 3 types of iris nodules found in the pediatric population:

- Lisch nodules
- Rushfield spots
- JXG nodules

Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules...

Mammillations? Aren't those a CNS thingamajig? You're thinking of the mammillary bodies, paired structures that are part of the limbic system.

OK, then what are iris mammillations?

Tiny pigmented nodules which, when present, are found in vast numbers diffusely scattered across the iris surface. Are they unilateral, or bilateral? Usually unilateral, but bilaterality occurs frequently enough that it can't be used to rule them out.

Are they associated with NF1? Yes (albeit not nearly as strongly as Lisch nodules).

Another sort of iris nodule needs mentioning here, because it is in the DDx for Lisch nodules. What is it?

In addition to NF1, iris mammillations have another important association. What is it?

Oculodermal melanocytosis, aka nevus of Ota and oculodermal melanocytosis.

What is oculodermal melanocytosis/nevus of Ota?

Ocular melanocytosis + hyperpigmentation of the ipsilateral periorbital skin.

What racial group is most likely to be affected?

Asians.

What is ocular melanocytosis?

A pigmented lesion of the episclera.

At what age does it present?

It is congenital, so birth.

Who gets it?

People of pigmented ethnicity, ie, blacks, Hispanics, Asians.

Is it unilateral, or bilateral?

Most (~95%) cases are unilateral.

Is it associated with ocular morbidity?

Yes, 10% of cases develop ipsilateral glaucoma.

What percent of ocular melanocytosis cases have oculodermal melanocytosis?

About half.

Does ocular melanocytosis possess malignant potential?

Only in whites.
Name 3 types of iris nodules found in the pediatric population:

- Lisch nodules
- Bensusan spots
- JXG nodules

Iris mammillations? Aren't those a CNS thingamajig?

Think of President Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules...

What is ocular melanocytosis?
A pigmented lesion of the episclera

At what age does it present?
It is congenital, so birth

Who gets it?
People of pigmented ethnicity, i.e., blacks, Hispanics, Asians

Is it unilateral, or bilateral?
Most (~95%) cases are unilateral

Is it associated with ocular morbidity?
Yes, 10% of cases develop ipsilateral glaucoma

What percent of ocular melanocytosis cases have oculodermal melanocytosis?
About half

Does ocular melanocytosis possess malignant potential?
Only in whites
Name 3 types of iris nodules found in the pediatric population:

- Lisch nodules
- Rushfield spots
- JXG nodules

Iris mammillations? Aren't those a CNS thingamajig? You're thinking of the **mammillary bodies**, paired structures that are part of the limbic system.

OK, then what are iris mammillations? Tiny pigmented nodules which, when present, are found in vast numbers diffusely scattered across the iris surface. Are they unilateral, or bilateral? Usually unilateral, but bilaterality occurs frequently enough that it can't be used to rule them out.

Are they associated with NF1? Yes (albeit not nearly as strongly as Lisch nodules).

Another sort of iris nodule needs mentioning here, because it is in the DDx for Lisch nodules. What is it? In addition to NF1, iris mammillations have another important association. What is it? Oculodermal melanocytosis, aka nevus of Ota and...oculodermal melanocytosis.

What is oculodermal melanocytosis/nevus of Ota? Ocular melanocytosis + hyperpigmentation of the ipsilateral periorbital skin.

What racial group is most likely to be affected? Asians.

What is ocular melanocytosis? A pigmented lesion of the episclera.

At what age does it present? It is congenital, so birth.

Who gets it? People of pigmented ethnicity, ie, blacks, Hispanics, Asians.

Is it unilateral, or bilateral? Most (~95%) cases are unilateral.

Is it associated with ocular morbidity? Yes, 10% of cases develop ipsilateral glaucoma.

What percent of ocular melanocytosis cases have oculodermal melanocytosis? About half.

Does ocular melanocytosis possess malignant potential? Only in whites.

Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules...
A

Name 3 types of iris nodules found in the pediatric population:

- Lisch nodules
- Rushfield spots
- JXG nodules
- Iris mammillations

Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules...

Mammillations? Aren’t those a CNS thingamajig? You’re thinking of the mammillary bodies, paired structures that are part of the limbic system.

OK, then what are iris mammillations?

Tiny pigmented nodules which, when present, are found in vast numbers diffusely scattered across the iris surface.

Are they unilateral, or bilateral?

Usually unilateral, but bilaterality occurs frequently enough that it can’t be used to rule them out.

Are they associated with NF1?

Yes (albeit not nearly as strongly as Lisch nodules).

Another sort of iris nodule needs mentioning here, because it is in the DDx for Lisch nodules. What is it?

In addition to NF1, iris mammillations have another important association. What is it?

Oculodermal melanocytosis, aka nevus of Ota and oculodermal melanocytosis.

What is oculodermal melanocytosis/nevus of Ota?

Ocular melanocytosis + hyperpigmentation of the ipsilateral periorbital skin.

What racial group is most likely to be affected?

Asians.

What is ocular melanocytosis?

A pigmented lesion of the episclera.

At what age does it present?

It is congenital, so birth.

Who gets it?

People of pigmented ethnicity, ie, blacks, Hispanics, Asians.

Is it unilateral, or bilateral?

Most (~95%) cases are unilateral.

Is it associated with ocular morbidity?

Yes, 10% of cases develop ipsilateral glaucoma.

What percent of ocular melanocytosis cases have oculodermal melanocytosis?

About half.

Does ocular melanocytosis possess malignant potential?

Only in whites.
Name 3 types of iris nodules found in the pediatric population:

- Lisch nodules
- Rushfield spots
- JXG nodules

Iris mammillations? Aren't those a CNS thingamajig?

You're thinking of the mammillary bodies, paired structures that are part of the limbic system. OK, then what are iris mammillations?

Tiny pigmented nodules which, when present, are found in vast numbers diffusely scattered across the iris surface. Are they unilateral, or bilateral? Usually unilateral, but bilaterality occurs frequently enough that it can't be used to rule them out.

Are they associated with NF1? Yes (albeit not nearly as strongly as Lisch nodules).

Another sort of iris nodule needs mentioning here, because it is in the DDx for Lisch nodules. What is it? In addition to NF1, iris mammillations have another important association. What is it? Oculodermal melanocytosis, aka nevus of Ota.

What is oculodermal melanocytosis/nevus of Ota? Ocular melanocytosis + hyperpigmentation of the ipsilateral periorbital skin. What racial group is most likely to be affected? Asians.

What is ocular melanocytosis? A pigmented lesion of the episclera. At what age does it present? It is congenital, so birth. Who gets it? People of pigmented ethnicity, ie, blacks, Hispanics, Asians.

Is it unilateral, or bilateral? Most (~95%) cases are unilateral. Is it associated with ocular morbidity? Yes, 10% of cases develop ipsilateral glaucoma.

What percent of ocular melanocytosis cases have oculodermal melanocytosis? About half.

Does ocular melanocytosis possess malignant potential? Only in whites. So, this episcleral lesion will transform into a melanoma?

Actually, no. In ocular (and oculodermal) melanocytosis, the entire uveal tract is overly melanotic. And it is the uvea that bears the increased risk of melanoma in white patients with melanocytosis. (Although white melanocytosis pts are also at increased risk of melanoma of the skin, conj and orbit as well.)

Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules...
Name 3 types of iris nodules found in the pediatric population:

- **Lisch nodules**
- **Birchfield spots**
- **JXG nodules**
- **Iris mammillations**

Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules...

Mammillations? Aren't those a CNS thingamajig?
You're thinking of the mammillary bodies, paired structures that are part of the limbic system.

OK, then what are iris mammillations?
Tiny pigmented nodules which, when present, are found in vast numbers diffusely scattered across the iris surface.

Are they unilateral, or bilateral?
Usually unilateral, but bilaterality occurs frequently enough that it can't be used to rule them out.

Are they associated with NF1?
Yes (albeit not nearly as strongly as Lisch nodules).

Another sort of iris nodule needs mentioning here, because it is in the DDx for Lisch nodules. What is it?
In addition to NF1, iris mammillations have another important association. What is it?
Oculodermal melanocytosis, aka nevus of Ota.

What is oculodermal melanocytosis/nevus of Ota?
Ocular melanocytosis + hyperpigmentation of the ipsilateral periorbital skin.

What racial group is most likely to be affected?
Asians.

What is ocular melanocytosis?
A pigmented lesion of the episclera.

At what age does it present?
It is congenital, so birth.

Who gets it?
People of pigmented ethnicity, ie, blacks, Hispanics, Asians.

Is it unilateral, or bilateral?
Most (~95%) cases are unilateral.

Is it associated with ocular morbidity?
Yes, 10% of cases develop ipsilateral glaucoma.

What percent of ocular melanocytosis cases have oculo-dermal melanocytosis?
About half.

Does ocular melanocytosis possess malignant potential?
Only in whites.

So, this episcleral lesion will transform into a melanoma?
Actually, no. In ocular (and oculodermal) melanocytosis, the entire uveal tract is overly melanotic. And it is the uvea that bears the increased risk of melanoma in white patients with melanocytosis. (Although white melanocytosis pts are also at increased risk of melanoma of the skin, conj and orbit as well.)
Name 3 types of iris nodules found in the pediatric population:

- Lisch nodules
- Rushfield spots
- JXG nodules
- Iris mammillations

Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules…

**What is ocular melanocytosis?**
A pigmented lesion of the episclera

At what age does it present?
It is congenital, so birth

Who gets it?
People of pigmented ethnicity, ie, blacks, Hispanics, Asians

What is the baseline risk of uveal melanoma in whites?
About 6 per million

What is the increased risk of melanoma in white patients with melanocytosis?
About half

Does ocular melanocytosis possess malignant potential?
Only in whites

What is oculodermal melanocytosis?
Ocular melanocytosis + hyperpigmentation of the ipsilateral periorbital skin

What is the risk in white folk with melanocytosis?
An astonishing 1 in 400!
Name 3 types of iris nodules found in the pediatric population:

- Lisch nodules
- Rushfield spots
- JXG nodules
- Iris mammillations

Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules…

Mammillations? Aren’t those a CNS thingamajig? You’re thinking of the mammillary bodies, paired nuclei of the thalamus that lie between the central gray matter and the cerebral cortex, are usually bilaterally symmetric and have a surface convoluted into a series of mamillary crests. OK, then what are iris mammillations?

Tiny pigmented nodules which, when present, are found in vast numbers diffusely scattered across the iris surface. Are they unilateral, or bilateral? Usually unilateral, but bilaterality occurs frequently enough that it can’t be used to rule them out. Are they associated with NF1? Yes (albeit not nearly as strongly as Lisch nodules).

Another sort of iris nodule needs mentioning here, because it is in the DDx for Lisch nodules. What is it? In addition to NF1, iris mammillations have another important association. What is it? Oculodermal melanocytosis, aka nevus of Ota and ocular melanocytosis.

What is oculodermal melanocytosis/nevus of Ota? Ocular melanocytosis + hyperpigmentation of the ipsilateral periorbital skin.

What racial group is most likely to be affected? Asians.

What is ocular melanocytosis? A pigmented lesion of the episclera. At what age does it present? It is congenital, so birth. Who gets it? People of pigmented ethnicity, ie, blacks, Hispanics, Asians. Is it unilateral, or bilateral? Most cases are unilateral. Is it associated with ocular morbidity? Yes, 10% of cases develop ipsilateral glaucoma. What percent of ocular melanocytosis cases have ocular dermal melanocytosis? About half. Does ocular melanocytosis possess malignant potential? Only in whites.

So, this episcleral lesion will transform into a melanoma? Actually, no. In ocular (and oculodermal) melanocytosis, the entire uveal tract is overly melanotic. And it is the uvea that bears the increased risk of melanoma in white patients with melanocytosis. (Although white melanocytosis pts are also at increased risk of melanoma of the skin, conj and orbit as well.)

What is the baseline risk of uveal melanoma in whites? About 6 per million. What is the risk in white folk with melanocytosis? An astonishing 1 in 400!
Name 3 types of iris nodules found in the pediatric population

- Lisch nodules
- Rushfield spots
- JXG nodules

Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules...

Mammillations? Aren’t those a CNS thingamajig? You’re thinking of the mammillary bodies, paired structures that are part of the limbic system.

OK, then what are iris mammillations?

Tiny pigmented nodules which, when present, are found in vast numbers diffusely scattered across the iris surface. Are they unilateral, or bilateral? Usually unilateral, but bilaterality occurs frequently enough that it can’t be used to rule them out.

Are they associated with NF1? Yes (albeit not nearly as strongly as Lisch nodules).

Another sort of iris nodule needs mentioning here, because it is in the DDx for Lisch nodules. What is it? In addition to NF1, iris mammillations have another important association. What is it? Oculodermal melanocytosis, aka nevus of Ota and... oculodermal melanocytosis.

What is oculodermal melanocytosis/nevus of Ota?

Ocular melanocytosis + hyperpigmentation of the ipsilateral periorbital skin.

What racial group is most likely to be affected? Asians.

What is ocular melanocytosis?

A pigmented lesion of the episclera. At what age does it present? It is congenital, so birth. Who gets it? People of pigmented ethnicity, ie, blacks, Hispanics, Asians. Is it unilateral, or bilateral? Most cases are unilateral. Is it associated with ocular morbidity? Yes, 10% of cases develop ipsilateral glaucoma.

What percent of ocular melanocytosis cases have oculo-dermal melanocytosis? About half.

Does ocular melanocytosis possess malignant potential? Only in whites. So, this episcleral lesion will transform into a melanoma? Actually, no. In ocular (and oculodermal) melanocytosis, the entire uveal tract is overly melanotic. And it is the uvea that bears the increased risk of melanoma in white patients with melanocytosis. (Although white melanocytosis pts are also at increased risk of melanoma of the skin, conj and orbit as well.)

What is the baseline risk of uveal melanoma in whites? About 6 per million. What is the risk in white folk with melanocytosis? An astonishing 1 in 400!
Name 3 types of iris nodules found in the pediatric population:

- Lisch nodules
- Rushfield spots
- JXG nodules

Think of President Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules...

What is ocular melanocytosis?
A pigmented lesion of the episclera

At what age does it present?
It is congenital, so birth

Who gets it?
People of pigmented ethnicity, i.e., blacks, Hispanics, Asians

What is the baseline risk of uveal melanoma in whites?
About 6 per million

What is the risk in white folk with melanocytosis?
An astonishing 1 in 400!

Another sort of iris nodule needs mentioning here, because it is in the DDx for Lisch nodules.

What is it?
Ocelodermal melanocytosis, aka nevus of Ota and oculodermal melanocytosis

What is oculodermal melanocytosis/nevus of Ota?
Ocular melanocytosis + hyperpigmentation of the ipsilateral periorbital skin

What racial group is most likely to be affected?
Asians

What is ocular melanocytosis?
A pigmented lesion of the episclera

At what age does it present?
It is congenital, so birth

Who gets it?
People of pigmented ethnicity, i.e., blacks, Hispanics, Asians

Is it unilateral, or bilateral?
Most cases are unilateral

Is it associated with NF1?
Yes (albeit not nearly as strongly as Lisch nodules)

Another sort of iris nodule needs mentioning here, because it is in the DDx for Lisch nodules.

What is it?
Ocellodermal melanocytosis, aka nevus of Ota and oculodermal melanocytosis

What is oculodermal melanocytosis/nevus of Ota?
Ocular melanocytosis + hyperpigmentation of the ipsilateral periorbital skin

What racial group is most likely to be affected?
Asians

What is ocular melanocytosis?
A pigmented lesion of the episclera

At what age does it present?
It is congenital, so birth

Who gets it?
People of pigmented ethnicity, i.e., blacks, Hispanics, Asians

Is it unilateral, or bilateral?
Most cases are unilateral

Is it associated with NF1?
Yes (albeit not nearly as strongly as Lisch nodules)

Mammillations? Aren’t those a CNS thingamajig? You’re thinking of the mammillary bodies, paired

What is the baseline risk of uveal melanoma in whites?
About 6 per million

What is the risk in white folk with melanocytosis?
An astonishing 1 in 400!

So, this episcleral lesion will transform into a melanoma?
Actually, no. In ocular (and oculodermal) melanocytosis, the entire uveal tract is overly melanotic. And it is the uvea that bears the increased risk of melanoma in white patients with melanocytosis.

What is the baseline risk of uveal melanoma in whites?
About 6 per million

What is the risk in white folk with melanocytosis?
An astonishing 1 in 400!

So, this episcleral lesion will transform into a melanoma?
Actually, no. In ocular (and oculodermal) melanocytosis, the entire uveal tract is overly melanotic. And it is the uvea that bears the increased risk of melanoma in white patients with melanocytosis.

What is the baseline risk of uveal melanoma in whites?
About 6 per million

What is the risk in white folk with melanocytosis?
An astonishing 1 in 400!
Name 3 types of iris nodules found in the pediatric population:

- Lisch nodules
- Brushfield spots
- JXG nodules

Brushfield spots are most strongly associated with what congenital condition?

Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules…
Name 3 types of iris nodules found in the pediatric population:

- **Lisch nodules**
- **Brushfield spots**
- **JXG nodules**

**Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules…**

*Brushfield spots are most strongly associated with what congenital condition?*

- Down syndrome
Name 3 types of iris nodules found in the pediatric population:

- **L**isch nodules
- **B**rushfield spots
- **J**XG nodules

*Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules...*
Name 3 types of iris nodules found in the pediatric population:

- **Lisch nodules**
- **Brushfield spots**
- **JXG nodules**

**Brushfield spots** are most strongly associated with what congenital condition?
- Down syndrome

What is the prevalence of Brushfield spots in the Down population?
- At least 90%

Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules…
Name 3 types of iris nodules found in the pediatric population:

- **Lisch nodules**
- **Brushfield spots**
- **JXG nodules**

*Brushfield spots are most strongly associated with what congenital condition?*
- Down syndrome

*What is the prevalence of Brushfield spots in the Down population?*
- At least 90%

*What is the clinical significance of Brushfield spots?*

*Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules…*
Name 3 types of iris nodules found in the pediatric population:

- **Lisch nodules**
- **Brushfield spots**
- **JXG nodules**

Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules…

**Brushfield spots** are most strongly associated with what congenital condition? Down syndrome

What is the prevalence of Brushfield spots in the Down population? At least 90%

What is the clinical significance of Brushfield spots? They have none
Name 3 types of iris nodules found in the pediatric population:

- **L**isch nodules
- **B**rushfield spots
- **J**XG nodules

Brushfield spots are most strongly associated with what congenital condition?
- **Down syndrome**

What is the prevalence of Brushfield spots in the Down population?
- At least 90%

What is the clinical significance of Brushfield spots?
- They have none

When a clinically identical iris finding occurs in a non-Down individual, what are the lesions called?

Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules…
Name 3 types of iris nodules found in the pediatric population:

- **Lisch nodules**
- **Brushfield spots**
- **JXG nodules**

---

Brushfield spots are most strongly associated with what congenital condition?

- Down syndrome

What is the prevalence of Brushfield spots in the Down population?

- At least 90%

What is the clinical significance of Brushfield spots?

- They have none

When a clinically identical iris finding occurs in a non-Down individual, what are the lesions called?

- Wolflin nodules

---

Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules…
Name 3 types of iris nodules found in the pediatric population:

- Lisch nodules
- Brushfield spots
- JXG nodules

Brushfield spots are most strongly associated with what congenital condition? Down syndrome

What is the prevalence of Brushfield spots in the Down population? At least 90%

Are Wolfflin nodules a common finding? Yes— they’re estimated to be present in about 25% of the population

When a clinically identical iris finding occurs in a non-Down individual, what are the lesions called? Wolfflin nodules

Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules…
Q/A

- Name 3 types of iris nodules found in the pediatric population:
  - Lisch nodules
  - Brushfield spots
  - JXG nodules

*Brushfield spots are most strongly associated with what congenital condition?*
- Down syndrome

*What is the prevalence of Brushfield spots in the Down population?*
- At least 90%

*When a clinically identical iris finding occurs in a non-Down individual, what are the lesions called?*
- Wolfflin nodules

*Are Wolfflin nodules a common finding?*
- Yes—they’re estimated to be present in about 25% of the population

*Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules…*
Name 3 types of iris nodules found in the pediatric population:

- Lisch nodules
- Brushfield spots
- JXG nodules

Brushfield spots are most strongly associated with what congenital condition? Down syndrome

What is the prevalence of Brushfield spots in the Down population? At least 90%

Are Wolfflin nodules a common finding? Yes--they’re estimated to be present in about 25% of the population

When a clinically identical iris finding occurs in a non-Down individual, what are the lesions called? Wolfflin nodules

Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules…
- Name 3 types of iris nodules found in the pediatric population:
  - Lisch nodules
  - Brushfield spots
  - JXG nodules

Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules...

What does JXG stand for?

Juvenile xanthogranuloma

In three words, what sort of condition is it?

It is a nonneoplastic histiocytic proliferation

What are the two hallmarks of JXG histology?

The presence of...

- Touton giant cells
- 'foamy macrophages'

At what age does JXG present?

The majority before age 1 year, and almost all by age 2

How does JXG usually present? (Hint: It's not ophthalmic)

As skin papules

When iris JXG nodules are present, is it uni-, or bilaterally?

Unilaterally

In what three ways are the iris nodules clinically significant?

--They are prone to spontaneous bleeding, with subsequent hyphema and secondary glaucoma--
--They are in the DDx as a 'masquerade syndrome' in peds uveitis--
--If enough are present, they will cause heterochromia iridis

Should they be removed surgically?

Only if the glaucoma is uncontrollable

What is the natural history of the disease?

JXG is self-limited, usually resolving by age 5 years
Name 3 types of iris nodules found in the pediatric population:

- **Lisch nodules**
- **Brushfield spots**
- **JXG nodules**

Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules…

What does JXG stand for? Juvenile xanthogranuloma

In three words, what sort of condition is it? **nonneoplastic histiocytic proliferation**

What are the two hallmarks of JXG histology?
- Touton giant cells
- 'foamy macrophages'

At what age does JXG present? The majority before age 1 year, and almost all by age 2

How does JXG usually present? (Hint: It's not ophthalmic) As skin papules

When iris JXG nodules are present, is it uni-, or bilaterally? Unilaterally

In what three ways are the iris nodules clinically significant?
- They are prone to spontaneous bleeding, with subsequent hyphema and secondary glaucoma
- They are in the DDx as a 'masquerade syndrome' in peds uveitis
- If enough are present, they will cause heterochromia iridis

Should they be removed surgically? Only if the glaucoma is uncontrollable

What is the natural history of the disease? JXG is self-limited, usually resolving by age 5 years
What does JXG stand for?
Juvenile xanthogranuloma

In three words, what sort of condition is it?

- Name 3 types of iris nodules found in the pediatric population:
  - Lisch nodules
  - Brushfield spots
  - JXG nodules

Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules...

What does JXG stand for?
Juvenile xanthogranuloma

In three words, what sort of condition is it?

- The presence of Touton giant cells
- The presence of 'foamy macrophages'

At what age does JXG present?
The majority before age 1 year, and almost all by age 2

How does JXG usually present? (Hint: It's not ophthalmic)
As skin papules

When iris JXG nodules are present, is it uni-, or bilaterally?
Unilaterally

In what three ways are the iris nodules clinically significant?
-- They are prone to spontaneous bleeding, with subsequent hyphema and secondary glaucoma
-- They are in the DDx as a 'masquerade syndrome' in peds uveitis
-- If enough are present, they will cause heterochromia iridis

Should they be removed surgically?
Only if the glaucoma is uncontrollable

What is the natural history of the disease?
JXG is self-limited, usually resolving by age 5 years
Name 3 types of iris nodules found in the pediatric population:
- Lisch nodules
- Brushfield spots
- JXG nodules

Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules.

What does JXG stand for?
Juvenile xanthogranuloma

In three words, what sort of condition is it?
It is a nonneoplastic histiocytic proliferation

What are the two hallmarks of JXG histology?
The presence of:
- Touton giant cells
- 'foamy macrophages'

At what age does JXG present?
The majority before age 1 year, and almost all by age 2

How does JXG usually present? (Hint: It's not ophthalmic)
As skin papules

When iris JXG nodules are present, is it uni-, or bilaterally?
Unilaterally

In what three ways are the iris nodules clinically significant?
- They are prone to spontaneous bleeding, with subsequent hyphema and secondary glaucoma
- They are in the DDx as a 'masquerade syndrome' in peds uveitis
- If enough are present, they will cause heterochromia iridis

Should they be removed surgically?
Only if the glaucoma is uncontrollable

What is the natural history of the disease?
JXG is self-limited, usually resolving by age 5 years
Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules...

What does JXG stand for?
Juvenile xanthogranuloma

In three words, what sort of condition is it?
It is a nonneoplastic histiocytic proliferation

What are the two hallmarks of JXG histology?
The presence of...
The presence of...

At what age does JXG present?
The majority before age 1 year, and almost all by age 2

How does JXG usually present? (Hint: It's not ophthalmic)
As skin papules

When iris JXG nodules are present, is it uni-, or bilaterally?
Unilaterally

In what three ways are the iris nodules clinically significant?
--They are prone to spontaneous bleeding, with subsequent hyphema and secondary glaucoma--They are in the DDx as a 'masquerade syndrome' in peds uveitis--If enough are present, they will cause heterochromia iridis

Should they be removed surgically?
Only if the glaucoma is uncontrollable

What is the natural history of the disease?
JXG is self-limited, usually resolving by age 5 years

Name 3 types of iris nodules found in the pediatric population:

- **L**isch nodules
- **B**rushfield spots
- **JXG** nodules
A

- Name 3 types of iris nodules found in the pediatric population:
  - Lisch nodules
  - Brushfield spots
  - JXG nodules

Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules...

What does JXG stand for?
Juvenile xanthogranuloma

In three words, what sort of condition is it?
It is a nonneoplastic histiocytic proliferation

What are the two hallmarks of JXG histology?
The presence of… Touton giant cells
The presence of… ‘foamy macrophages’

At what age does JXG present?
The majority before age 1 year, and almost all by age 2

How does JXG usually present? (Hint: It's not ophthalmic)
As skin papules

When iris JXG nodules are present, is it uni-, or bilaterally?
Unilaterally

In what three ways are the iris nodules clinically significant?
-- They are prone to spontaneous bleeding, with subsequent hyphema and secondary glaucoma
-- They are in the DDx as a ‘masquerade syndrome’ in pediatrics uveitis
-- If enough are present, they will cause heterochromia iridis

Should they be removed surgically?
Only if the glaucoma is uncontrollable

What is the natural history of the disease?
JXG is self-limited, usually resolving by age 5 years
Q

Name 3 types of iris nodules found in the pediatric population:
- Lisch nodules
- Brushfield spots
- JXG nodules

Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules...

What does JXG stand for?
Juvenile xanthogranuloma

In three words, what sort of condition is it?
It is a nonneoplastic histiocytic proliferation

What are the two hallmarks of JXG histology?
The presence of...Touton giant cells
The presence of...'foamy macrophages'

At what age does JXG present?
The majority before age 1 year, and almost all by age 2

How does JXG usually present? (Hint: It's not ophthalmic)
As skin papules

When iris JXG nodules are present, is it uni-, or bilaterally?
Unilaterally

In what three ways are the iris nodules clinically significant?
--They are prone to spontaneous bleeding, with subsequent hyphema and secondary glaucoma--They are in the DDx as a 'masquerade syndrome' in peds uveitis--If enough are present, they will cause heterochromia iridis

Should they be removed surgically?
Only if the glaucoma is uncontrollable

What is the natural history of the disease?
JXG is self-limited, usually resolving by age 5 years
Name 3 types of iris nodules found in the pediatric population:

- **L**isch nodules
- **B**rushfield spots
- **JXG** nodules

Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules...

**What does JXG stand for?**
Juvenile xanthogranuloma

**In three words, what sort of condition is it?**
It is a nonneoplastic histiocytic proliferation

**What are the two hallmarks of JXG histology?**
The presence of… **Touton giant cells**
The presence of… ’foamy macrophages’

**At what age does JXG present?**
The majority before age 1 year, and almost all by age 2

**What is the natural history of the disease?**
JXG is self-limited, usually resolving by age 5 years
Name 3 types of iris nodules found in the pediatric population:

- **Lisch nodules**
- **Brushfield spots**
- **JXG nodules**

Think of President Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules...

What does JXG stand for?  
Juvenile xanthogranuloma

In three words, what sort of condition is it?  
It is a **nonneoplastic histiocytic proliferation**

What are the two hallmarks of JXG histology?  
The presence of… **Touton giant cells**  
The presence of… **'foamy macrophages'**

At what age does JXG present?  
The majority before age 1 year, and almost all by age 2

How does JXG usually present?  
(Hint: It’s not ophthalmic)

What is the natural history of the disease?  
JXG is self-limited, usually resolving by age 5 years
A

- Name 3 types of iris nodules found in the pediatric population
  - Lisch nodules
  - Brushfield spots
  - JXG nodules

Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules...
Name 3 types of iris nodules found in the pediatric population:

- **Lisch nodules**
- **Brushfield spots**
- **JXG nodules**

Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules…

**What does JXG stand for?**
Juvenile xanthogranuloma

**In three words, what sort of condition is it?**
It is a nonneoplastic histiocytic proliferation

**What are the two hallmarks of JXG histology?**
The presence of… **Touton giant cells**
The presence of… ‘**foamy macrophages**’

**At what age does JXG present?**
The majority before age 1 year, and almost all by age 2

**How does JXG usually present? (Hint: It’s not ophthalmic)**
As skin papules

**When iris JXG nodules are present, is it uni-, or bilaterally?**
Unilaterally

**In what three ways are the iris nodules clinically significant?**
-- They are prone to spontaneous bleeding, with subsequent hyphema and secondary glaucoma
-- They are in the DDx as a 'masquerade syndrome' in peds uveitis
-- If enough are present, they will cause heterochromia iridis

**Should they be removed surgically?**
Only if the glaucoma is uncontrollable

**What is the natural history of the disease?**
JXG is self-limited, usually resolving by age 5 years.
A

- Name 3 types of iris nodules found in the pediatric population
  - Lisch nodules
  - Brushfield spots
  - JXG nodules

Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules...

What does JXG stand for?
Juvenile xanthogranuloma

In three words, what sort of condition is it?
It is a nonneoplastic histiocytic proliferation

What are the two hallmarks of JXG histology?
The presence of... Touton giant cells
The presence of... 'foamy macrophages'

At what age does JXG present?
The majority before age 1 year, and almost all by age 2

How does JXG usually present? (Hint: It’s not ophthalmic)
As skin papules

When iris JXG nodules are present, is it uni-, or bilaterally?
Unilaterally

What is the natural history of the disease?
JXG is self-limited, usually resolving by age 5 years
Name 3 types of iris nodules found in the pediatric population:

- **Lisch nodules**
- **Brushfield spots**
- **JXG nodules**

Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules...

**What does JXG stand for?**
Juvenile xanthogranuloma

**In three words, what sort of condition is it?**
It is a *nonneoplastic histiocytic proliferation*

**What are the two hallmarks of JXG histology?**
The presence of… **Touton giant cells**
The presence of… ‘**foamy macrophages**’

**At what age does JXG present?**
The majority before age 1 year, and almost all by age 2

**How does JXG usually present?** (Hint: It’s not ophthalmic)
As skin papules

**When iris JXG nodules are present, is it uni-, or bilaterally?**
Unilaterally

**In what three ways are the iris nodules clinically significant?**

- They are prone to spontaneous bleeding, with subsequent hyphema and secondary glaucoma
- They are in the DDx as a ‘masquerade syndrome’ in peds uveitis
- If enough are present, they will cause heterochromia iridis

Should they be removed surgically?
Only if the glaucoma is uncontrollable

**What is the natural history of the disease?**
JXG is self-limited, usually resolving by age 5 years
Lisch nodules

Brushfield spots

JXG nodules

What does JXG stand for?
Juvenile xanthogranuloma

In three words, what sort of condition is it?
It is a nonneoplastic histiocytic proliferation

What are the two hallmarks of JXG histology?
The presence of…

- Touton giant cells
- ‘foamy macrophages’

At what age does JXG present?
The majority before age 1 year, and almost all by age 2

How does JXG usually present? (Hint: It’s not ophthalmic)
As skin papules

When iris JXG nodules are present, is it uni-, or bilaterally?
Unilaterally

In what three ways are the iris nodules clinically significant?
--They are prone to spontaneous bleeding, with subsequent hyphema and secondary glaucoma
--They are in the DDx as a ‘masquerade syndrome’ in peds uveitis
--If enough are present, they will cause heterochromia iridis

Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules…
Name 3 types of iris nodules found in the pediatric population:

- **Lisch nodules**
- **Brushfield spots**
- **JXG nodules**

What does JXG stand for? Juvenile xanthogranuloma

In three words, what sort of condition is it? It is a **nonneoplastic histiocytic proliferation**

What are the two hallmarks of JXG histology? The presence of... **Touton giant cells** The presence of... **'foamy macrophages'**

At what age does JXG present? The majority before age 1 year, and almost all by age 2

How does JXG usually present? (Hint: It’s not ophthalmic) As skin papules

When iris JXG nodules are present, is it uni-, or bilaterally? Unilaterally

In what three ways are the iris nodules clinically significant? --They are prone to spontaneous bleeding, with subsequent hyphema and secondary glaucoma --They are in the DDx as a 'masquerade syndrome' in peds uveitis --If enough are present, they will cause heterochromia iridis

Should they be removed surgically?

*Think of president Lyndon Baines Johnson...*
Name 3 types of iris nodules found in the pediatric population:

- **Lisch nodules**
- **Brushfield spots**
- **JXG nodules**

*Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules…*

**What does JXG stand for?**
Juvenile xanthogranuloma

**In three words, what sort of condition is it?**
It is a *nonneoplastic histiocytic proliferation*

**What are the two hallmarks of JXG histology?**
The presence of… **Touton giant cells**
The presence of… **'foamy macrophages'**

**At what age does JXG present?**
The majority before age 1 year, and almost all by age 2

**How does JXG usually present? (Hint: It’s not ophthalmic)**
As skin papules

**When iris JXG nodules are present, is it uni-, or bilaterally?**
Unilaterally

**In what three ways are the iris nodules clinically significant?**
--- They are prone to spontaneous bleeding, with subsequent hyphema and secondary glaucoma
--- They are in the DDx as a ‘masquerade syndrome’ in peds uveitis
--- If enough are present, they will cause heterochromia iridis

**Should they be removed surgically?**
**Only** if the glaucoma is uncontrollable
What does JXG stand for?
Juvenile xanthogranuloma

In three words, what sort of condition is it?
It is a nonneoplastic histiocytic proliferation

What are the two hallmarks of JXG histology?
The presence of...Touton giant cells
The presence of...‘foamy macrophages’

At what age does JXG present?
The majority before age 1 year, and almost all by age 2

How does JXG usually present? (Hint: It’s not ophthalmic)
As skin papules

When iris JXG nodules are present, is it uni-, or bilaterally?
Unilaterally

In what three ways are the iris nodules clinically significant?
--They are prone to spontaneous bleeding, with subsequent hyphema and secondary glaucoma
--They are in the DDx as a ‘masquerade syndrome’ in peds uveitis
--If enough are present, they will cause heterochromia iridis

Should they be removed surgically?
Only if the glaucoma is uncontrollable

What is the natural history of the disease?
JXG is self-limited, usually resolving by age 5 years

Q

Name 3 types of iris nodules found in the pediatric population

- Lisch nodules
- Brushfield spots
- JXG nodules

Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules...
Name 3 types of iris nodules found in the pediatric population:

- **Lisch nodules**
- **Brushfield spots**
- **JXG nodules**

Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules...

What does JXG stand for?  
Juvenile xanthogranuloma

In three words, what sort of condition is it?  
It is a nonneoplastic histiocytic proliferation

What are the two hallmarks of JXG histology?  
The presence of… **Touton giant cells**  
The presence of… **‘foamy macrophages’**

At what age does JXG present?  
The majority before age 1 year, and almost all by age 2

How does JXG usually present? (Hint: It’s not ophthalmic)  
As skin papules

When iris JXG nodules are present, is it uni-, or bilaterally?  
Unilaterally

In what three ways are the iris nodules clinically significant?  
--They are prone to spontaneous bleeding, with subsequent hyphema and secondary glaucoma  
--They are in the DDx as a ‘masquerade syndrome’ in peds uveitis  
--If enough are present, they will cause heterochromia iridis

Should they be removed surgically?  
**Only** if the glaucoma is uncontrollable

What is the natural history of the disease?  
JXG is self-limited, usually resolving by age 5 years
Name 3 types of iris nodules found in the pediatric population:
- Lisch nodules
- Rushfield spots
- JXG nodules

Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules…

What does JXG stand for?
Juvenile xanthogranuloma

In three words, what sort of condition is it?
It is a nonneoplastic histiocytic proliferation

What are the two hallmarks of JXG histology?
The presence of…
- Touton giant cells
- *foamy macrophages*

At what age does JXG present?
The majority before age 1 year, and almost all by age 2

How does JXG usually present? (Hint: It’s not ophthalmic)
As skin papules

When iris JXG nodules are present, is it uni-, or bilaterally?
Unilaterally

In what three ways are the iris nodules clinically significant?
- They are prone to spontaneous bleeding, with subsequent hyphema and secondary glaucoma
- They are in the DDx as a 'masquerade syndrome' in peds uveitis
- If enough are present, they will cause heterochromia iridis

Should they be removed surgically?
Only if the glaucoma is uncontrollable

What is the natural history of the disease?
JXG is self-limited, usually resolving by age 5 years
Q

- Name 3 types of iris nodules found in the pediatric population:
  - Lisch nodules
  - Rushfield spots
  - JXG nodules

Think of President Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules…

What does JXG stand for?
Juvenile xanthogranuloma

In three words, what sort of condition is it?
It is a nonneoplastic histiocytic proliferation

What are the two hallmarks of JXG histology?
The presence of…
  - Touton giant cells
  - *foamy macrophages*

At what age does JXG present?
The majority before age 1 year, and almost all by age 2

How does JXG usually present? (Hint: It’s not ophthalmic)
As skin papules

When iris JXG nodules are present, is it uni-, or bilaterally?
Unilaterally

In what three ways are the iris nodules clinically significant?
-- They are prone to spontaneous bleeding, with subsequent hyphema and secondary glaucoma
-- They are in the DDx as a ‘masquerade syndrome’ in peds uveitis
-- If enough are present, they will cause heterochromia iridis

Should they be removed surgically?
Only if the glaucoma is uncontrollable

What is the natural history of the disease?
JXG is self-limited, usually resolving by age 5 years

Speaking of ‘foamy macrophages’…
What dz comes to mind if, instead of a young child with iris nodules, the pt in question was a middle-aged white guy with bilateral panuveitis?
And a hx of chronic migratory arthritis?  

--- Clue #2

--- They are prone to spontaneous bleeding, with subsequent hyphema and secondary glaucoma
--- They are in the DDx as a ‘masquerade syndrome’ in peds uveitis
--- If enough are present, they will cause heterochromia iridis

Should they be removed surgically?
Only if the glaucoma is uncontrollable

What is the natural history of the disease?
JXG is self-limited, usually resolving by age 5 years
Name 3 types of iris nodules found in the pediatric population:

- Lisch nodules
- Rushfield spots
- JXG nodules

Think of President Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules...

What does JXG stand for?
Juvenile xanthogranuloma

In three words, what sort of condition is it?
It is a nonneoplastic histiocytic proliferation

What are the two hallmarks of JXG histology?
The presence of...
- Touton giant cells
- *foamy macrophages*

Speaking of *foamy macrophages*…

What disease comes to mind if, instead of a young child with iris nodules, the pt in question was a middle-aged white guy with bilateral panuveitis?
And a hx of chronic migratory arthritis?
Associated with chronic diarrhea? — Need another?

---

They are prone to spontaneous bleeding, with subsequent hyphema and secondary glaucoma

They are in the DDx as a 'masquerade syndrome' in peds uveitis

If enough are present, they will cause heterochromia iridis

Should they be removed surgically?
Only if the glaucoma is uncontrollable

What is the natural history of the disease?
JXG is self-limited, usually resolving by age 5 years
Name 3 types of iris nodules found in the pediatric population:

- Lisch nodules
- Rushfield spots
- JXG nodules

Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules...

What does JXG stand for?
Juvenile xanthogranuloma

In three words, what sort of condition is it?
It is a nonneoplastic histiocytic proliferation

What are the two hallmarks of JXG histology?
The presence of... Touton giant cells
The presence of... *foamy macrophages*

At what age does JXG present?
The majority before age 1 year, and almost all by age 2

How does JXG usually present? (Hint: It's not ophthalmic)
As skin papules

When iris JXG nodules are present, is it unilaterally or bilaterally?
Unilaterally

In what three ways are the iris nodules clinically significant?
--They are prone to spontaneous bleeding, with subsequent hyphema and secondary glaucoma
--They are in the DDx as a 'masquerade syndrome' in peds uveitis
--If enough are present, they will cause heterochromia iridis

Should they be removed surgically?
Only if the glaucoma is uncontrollable

What is the natural history of the disease?
JXG is self-limited, usually resolving by age 5 years

Speaking of ‘foamy macrophages’...
What dz comes to mind if, instead of a young child with iris nodules, the pt in question was a middle-aged white guy with bilateral panuveitis?
And a hx of chronic migratory arthritis?
Associated with chronic diarrhea?
And CNS symptoms--seizures, dementia, coma? Last chance--answer is next!

Whipple's disease

"They are in the DDx as a 'masquerade syndrome' in peds uveitis"
Name 3 types of iris nodules found in the pediatric population:

- Lisch nodules
- Burchfield spots
- JXG nodules

Think of president Lyndon Baines Johnson (known colloquially as LBJ) as having iris nodules...

What does JXG stand for?
Juvenile xanthogranuloma

In three words, what sort of condition is it?
It is a nonneoplastic histiocytic proliferation

What are the two hallmarks of JXG histology?
The presence of...
- Touton giant cells
- *foamy macrophages*

At what age does JXG present?
The majority before age 1 year, and almost all by age 2

How does JXG usually present? (Hint: It's not ophthalmic)
As skin papules

When iris JXG nodules are present, is it uni-, or bilaterally?
Unilaterally

In what three ways are the iris nodules clinically significant?
--They are prone to spontaneous bleeding, with subsequent hyphema and secondary glaucoma
--They are in the DDx as a 'masquerade syndrome' in peds uveitis
--If enough are present, they will cause heterochromia iridis

Should they be removed surgically?
Only if the glaucoma is uncontrollable

What is the natural history of the disease?
JXG is self-limited, usually resolving by age 5 years

Speaking of ‘foamy macrophages’…
What dz comes to mind if, instead of a young child with iris nodules, the pt in question was a middle-aged white guy with bilateral panuveitis?
And a hx of chronic migratory arthritis?
Associated with chronic diarrhea?
And CNS symptoms--seizures, dementia, coma?
Whipple’s disease ← Ding ding ding

---
Congenital iris ectropion syndrome consists of ectropion uveae plus…
A

- *Congenital iris ectropion syndrome* consists of unilateral ectropion uveae plus...
Q

- **Congenital iris ectropion syndrome** consists of **unilateral** ectropion uveae plus...
  - ...iris **surface** abnormality--is it **glassy and cryptless**, or **extra corrugated**?
Congenital iris ectropion syndrome consists of unilateral ectropion uveae plus…

...iris surface abnormality--is it glassy and cryptless, or extra corrugated? Glassy and cryptless

A
Congenital iris ectropion syndrome consists of unilateral ectropion uveae plus…

- …iris **surface** abnormality--is it *glassy and cryptless*, or extra corrugated? **Glassy** and **cryptless**
- …iris **insertion** abnormality--**too high** or **too low**?
A

- Congenital iris ectropion syndrome consists of unilateral ectropion uveae plus...
  - ...iris surface abnormality--is it *glassy and cryptless*, or *extra corrugated*? *Glassy* and *cryptless*
  - ...iris insertion abnormality--*too high* or *too low*? *Too high*
Congenital iris ectropion syndrome consists of unilateral ectropion uveae plus…

- …iris surface abnormality--is it glassy and cryptless, or extra corrugated? **Glassy and cryptless**
- …iris insertion abnormality--**too high** or **too low**? **Too high**
- …**dysgenesis** of what anterior structure?
Congenital iris ectropion syndrome consists of unilateral ectropion uveae plus…

- …iris surface abnormality--is it glassy and cryptless, or extra corrugated? Glassy and cryptless
- …iris insertion abnormality--too high or too low? Too high
- …dysgenesis of what anterior structure? The TM (trabecular meshwork)
Congenital iris ectropion syndrome consists of unilateral ectropion uveae plus…

- ...iris surface abnormality--is it glassy and cryptless, or extra corrugated? Glassy and cryptless
- ...iris insertion abnormality--too high or too low? Too high
- ...dysgenesis of what anterior structure? The TM
- ...what blinding complication?
Congenital iris ectropion syndrome consists of unilateral ectropion uveae plus...

- ...iris surface abnormality--is it glassy and cryptless, or extra corrugated? Glassy and cryptless
- ...iris insertion abnormality--too high or too low? Too high
- ...dysgenesis of what anterior structure? The TM
- ...what blinding complication? Glaucoma
- **Congenital iris ectropion syndrome** consists of unilateral **ectropion uveae** plus...
  - ...iris surface abnormality--is it glassy and cryptless, or extra corrugated? **Glassy** and **cryptless**
  - ...iris insertion abnormality--too high or too low? Too **high**
  - ...**dysgenesis** of what anterior structure? The **TM**
  - ...what **blinding complication**? **Glaucoma**

- **What does ectropion uveae refer to, and why is the term technically a misnomer?**
Congenital iris ectropion syndrome consists of unilateral ectropion uveae plus...

- iris surface abnormality--is it glassy and cryptless, or extra corrugated? Glassy and cryptless
- iris insertion abnormality--too high or too low? Too high
- dysgenesis of what anterior structure? The TM
- what blinding complication? Glaucoma

What does ectropion uveae refer to, and why is the term technically a misnomer? Ectropion uveae refers to the presence of posterior pigmented iris epithelium on the anterior surface of the iris. It is a misnomer because the posterior pigmented epithelium derives from neuroectoderm, not uvea.

(I guess the term ectropion neuroectodermiae didn’t scan well…)