

INFANTILE EXOTROPIA

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INFANTILE XT

- Usage often imprecise
- Variation in definitions \approx number of investigators
- ? onset day 1 of life
- ? constant / intermittent
- Any / large angle?
- Associated systemic / ocular conditions included / excluded?

CORE SLIDE 1: 4 MAIN GROUPS

- 1. 'True' infantile XT: the mirror image of congenital ET
- 2. Early onset 'regular' intermittent XT [wch can later turn out to be constant XT]
- These [1 & 2] are often lumped together
- 3. Sensory problem – monocular cataract, RB
- 4. CNS problem
- 5. Late presentation of #1

CORE SLIDE 2

1. This is uncommon - to - rare
2. This is a **high pathology** condition
2. All pts need careful eye exam & repeated careful eye exams
3. Most / all need pediatrician or pediatric neurology assessment - esp. with optometric / ophthalmology referrals
4. Probable 'true' infantile XT: early surgery
5. Early onset intermittent XT: uncertain: Saunders vsHiles. Some may improve. Early surgery no worse than late surgery

CORE SLIDE 3

- Results : Constant XT in Y1 \approx intermittent XT
- Reoperation rate 20 - 40%
- 60- 100% end up $\leq 10 \Delta$
- 20-50% have some / good stereo.
Intermittent : up to 70 %
- 0-25% have amblyopia

4 MAIN GROUPS

- **1. 'True' infantile XT: mirror image of congenital ET**
- 2. Early onset 'regular' intermittent XT
- 3. Sensory problem – monocular cataract, RB
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'True' infantile XT

Older literature:

Early onset

Large angle

Newer literature:

Should have the cardinal features of congenital strabismus:

- SPA smooth pursuit asymmetry
- LMLN - H &/or T, and its sequelae inc DVD

Infantile exotropia in healthy children.

Rubin SE, Nelson LB, Wagner RS, Simon JW, Catalano RA

Ophthalmic Surg. 1988 Nov;19(11):792-4.

Long Island Jewish Medical Center & Wills

- 13 pts with large angle constant XT during Y1
- LR Rc OU in all. 2 pts : reoperation.
- Clinical characteristics and management of this unusual condition – ‘similar to infantile esotropia’.
- *1988: early onset and large angle were considered to be cardinal features.*
- *These pts were not known to have SPA, LMLN,....so some / many are NOT ‘true’ congenital XT, but ‘regular’ early onset XT that has become constant*

Hopkins series #1

- **1. 'True' infantile XT: mirror image of congenital ET**
- **2. Early onset 'regular' intermittent XT**
- **NOT:**
- **3. Sensory problem – monocular cataract, RB**
- **4. CNS problem**

Constant vs. intermittent XT in Y1

Long-term outcome of uncomplicated infantile exotropia.

[Hunter DG](#), [Kelly JB](#), [Buffenn AN](#), [Ellis FJ](#). J AAPOS. Dec 2001 Dec

Wilmer Institute, Johns Hopkins University School of Medicine

- **Exclude** : previous strab surgery, resolution by 3 mo, **concomitant systemic or ocular disease**.
- N=13. 6 constant , 7 intermittent. 12/13 had surgery
- Larger initial angle in the constant XT group.
- Av follow-up 5y
- Reoperation rate 27%
- 82% : final horizontal deviations $\leq 10 \Delta$
- Incidence of A/V patterns (38%), DVD (46%) & binocularity (70%) similar between groups
- **Half of infantile XT pts have intermittent XT; similar clinical outcomes to constant XT**

Infantile exotropia. [Biglan AW](#).....

JPOS. 1996 Mar-Apr;33(2):79-84.Pittsburgh

- 12 patients with XT $\geq 15 \Delta$ followed for $\geq 4y$.
- EXCLUDE: neurologic defects, prematurity, trauma, craniofacial syndromes, orbital abnormalities or ocular defects that would reduce vision.
- Mean age first exam 8 mo
- XT intermittent in 4, constant in 8.
- 3 had amblyopia.
- 10 had surgery.
- 4 : additional procedures, mostly to correct oblique OA & DVD.
- Most recent visit :
 1. all 12 pts had equal VA and satisfactory ocular alignment.
 2. 5 had fusion at distance and near ...2 had stereo ≥ 100 “.
- ...nystagmus is rare

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Sensory results after LR recession for intermittent XT operated <2y of age

- Saunders RA..... J AAPOS. 4/2008 South Carolina
Early surgery for early-onset intermittent XT is controversial –
 1. does not always progress
 2. postoperative ET has adverse consequences - suppression, amblyopia, and loss of binocular vision.
- .. 12 pts with intermittent XT, onset < 1y who had LR Rc OU before 2y & follow-up exams at ≥4y (Worth 4-Dot and Titmus stereo likely to be reliable)

Results after LR Rc for [XT] operated <2y n=12

- Stereo results:
- 40" in 2 pts, 100" in 3, 140-400" in 2, 0 in 5.
- 7 pts (58%) : phoria or intermittent tropia <10Δ at D & N.
- No pt required treatment for amblyopia.

BUT.....

- Arch Ophthalmol. 1968 Oct;80(4):436-42.
- Long-term observations on unoperated intermittent exotropia.
- **Hiles DA,Costenbader FD.**
- **Infants for whom XT surgery was recommended, but parents declined: 25% had no XT on followup**

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SENSORY PROBLEM

- **monocular cataract, RB,
unilateral myopia,
.....important visual and
systemic implications**

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HOPKINS SERIES #2

Prevalence of systemic and ocular disease in infantile exotropia: comparison with infantile esotropia. [HunterDG](#), [Ellis FJ](#). Ophthalmology. 1999

..prevalence of ocular disease & systemic illness in pts with XT in infancy.

- Chart review : 70 pts with XT in Y1 c.f. with 136 pts with ET before 1y

67% of XT and 49% of ET pts : coexisting ocular or systemic abnormality.

- Systemic disorders more frequent than ocular disorders in both the XT & ET

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Marseille cohort - all had MRI

Baeteman et alii J Fr Ophtalmol. 2008 Mar.

Primary exotropia: importance of cerebral MRI

- **Primary XT** presents on the first day of life and persists > 1y, n =47.
- MRI @ average age, 16 mo.

Marseille cohort - results

- Increased rate of abnormal MRI as angle of XT increased
- **~80% had a pathological ophthalmologic exam** (amblyopia, ptosis, head posture, Duane's syndrome, cataract, albinism, or pigmentary retinopathy).
- **~70 % MRIs abnormal** – of these 69% white matter injury, 40% gray matter injury, and 20% thin corpus callosum
- ~40% nystagmus
- ~40% optic nerve hypoplasia.
- ~60%: significant associated pathology - prematurity, fetal distress, plagiocephaly, psychomotor delay, epilepsy.
- **Only 3/47 had isolated exotropia**

Messages 1

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2. This is a **high pathology** condition
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4. Probable 'true' infantile XT: early surgery
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Messages 2

- Results of Constant XT \approx intermittent
- Reoperation rate 20 - 40%
- 60- 100% $\leq 10 \Delta$
- 20-50% have some / good stereo.
- Intermittent: 70 % have some / good stereo.
- 0-25% have amblyopia