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]
3. Sensory problem – monocular cataract, RB
4. CNS problem
5. Late presentation of #1
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 vs Hiles. Some may improve. Early surgery no worse than late surgery
CORE SLIDE 3

• Results: Constant XT in Y1 ≈ intermittent XT
• Reoperation rate 20 - 40%
• 60-100% end up ≤ 10 Δ
• 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
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• 5. Late presentation of #1
‘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
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 ≤ 10 Δ
• 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**

- 12 patients with XT ≥ 15 Δ followed for ≥ 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....... 


• 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
4 MAIN GROUPS

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SENSOR PROBLEM

• monocular cataract, RB, unilateral myopia,
......important visual and systemic implications
4 MAIN GROUPS

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..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

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. This is uncommon - to - rare
2. This is **a high pathology** condition
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4. Probable ‘true’ infantile XT: early surgery
5. Early onset intermittent XT: uncertain: Saunders vs Hiles. Some may improve. Early surgery no worse than late surgery
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