Eye movement recordings for childhood onset nystagmus:

When you don't need them

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Introduction

- **Eye movement recordings (EMR)** for nystagmus help identify features that guide **optical and surgical** management and offer **prognostic advice.**
- We send all patients with nystagmus for **EMR** as part of their initial assessment.
- EMR is not only useful in a **research setting and for descriptive purposes**, but is also an excellent tool for **diagnosing and managing nystagmus** prior to considering treatment options.

But is it always necessary to get EMR?

We try to identify the characteristics of nystagmus that may indicate when EMR may not be necessary.

Methods and Results:

- Retrospective review of 44 consecutive cases of nystagmus with EMR between October 2011 to Jan 2015

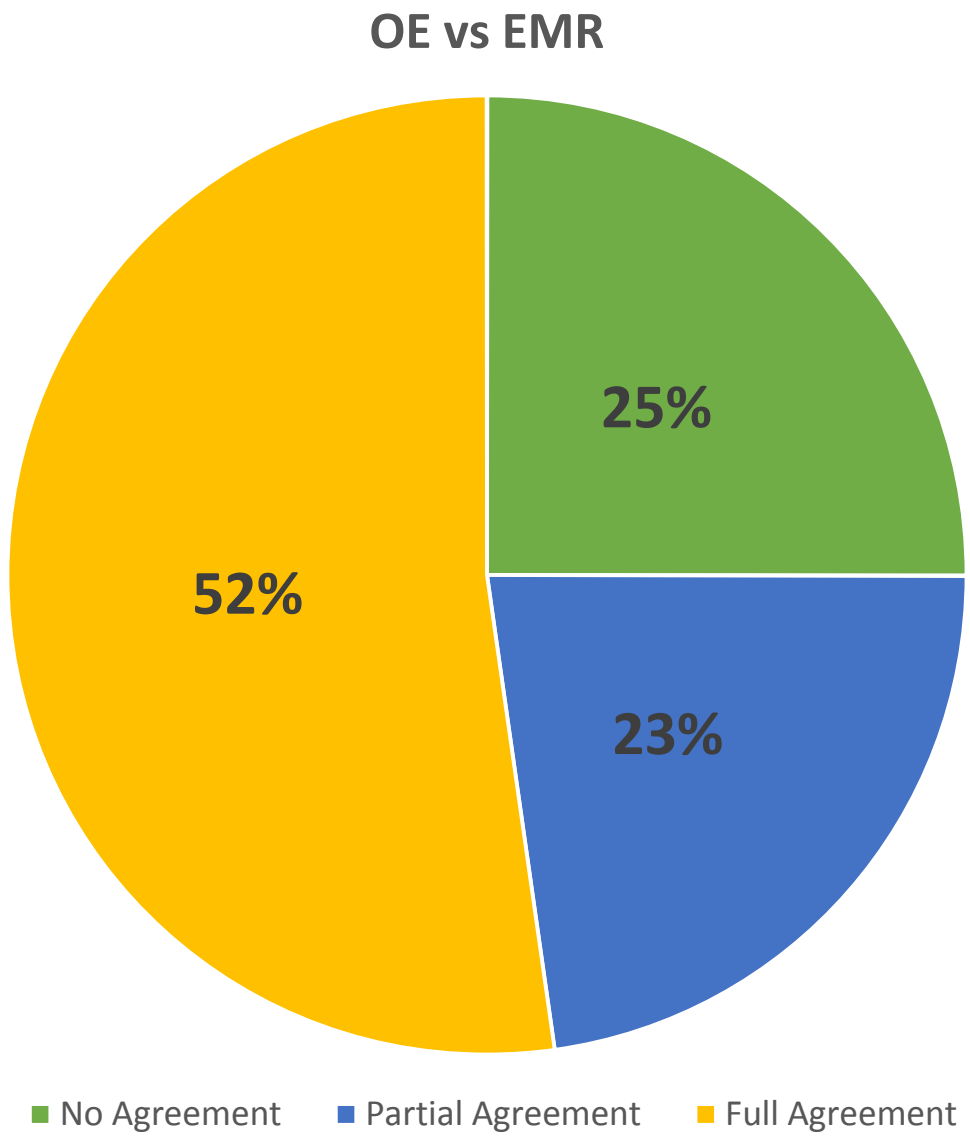
- The level of agreement between an **office examination (OE)** vs **EMR** in the diagnosis of common forms of childhood-onset nystagmus was assessed.

Results:

- **Total N: 44 [**Male**:** 28 (63%) Female = 16 (36%)]
- **Age when EMR performed:** 19.8 years ± 17.7 years
- **N with Abnormal Head Position (AHP)**= 34 (77.2%)
- **N with Neurological/ Developmental Abnormalities** = 7 (15.9%)
- Mostly Caucasian, middle class, neurologically and developmentally healthy
- Many of the ‘childhood’ cases 1st present as teens or adults and a few as old adults

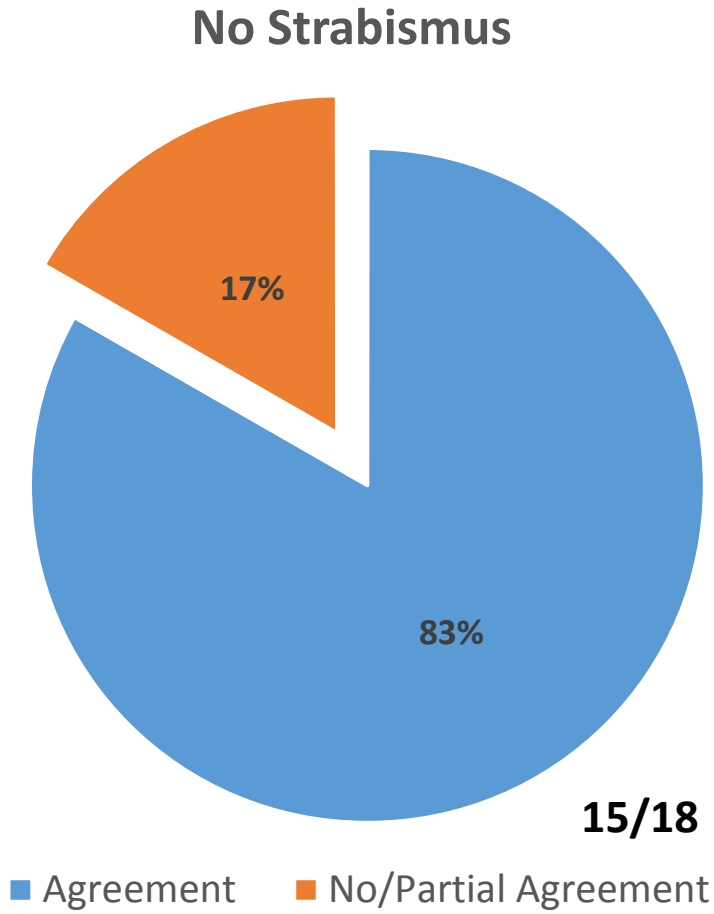
Overall Office Diagnosis vs EMR Diagnosis

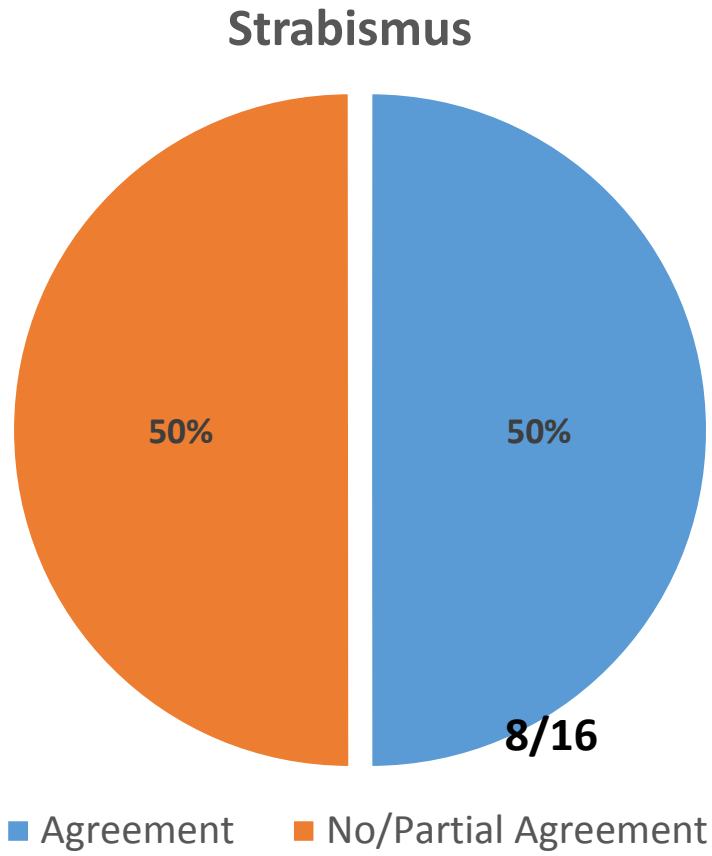
- **No agreement N=11 (25%)**
- Partial agreement N=10 (22.7%)
- **Agreement N=23 (52.2%)**



Strabismus- OE vs EMR

- There is a significantly higher level of agreement between OE and EMR when there is no strabismus. (**p =0.038)**





Infantile Nystagmus Syndrome (INS) Features assessed in office:

- Conjugate, **Horizontal Jerk Nystagmus**
- **Eccentric null**
- **For Distance viewing is ALWAYS Face turn (FT) on same side, never straight and never to same side**
- **Same FT with either eye fixing**
- **Convergence null for near, with little/no FT**
- **Convergence null for Distance.**
- **No strabismus**
- Increases with fixation attempt
- May have associated sensory defects (albinism, achromatopisia)

FT= Face Turn

Infantile Nystagmus Syndrome (INS) – example patient

With protractor, **Face Turn to Left 25+ °, tip up 20°**

7Δ BO OU & -1 DS OU

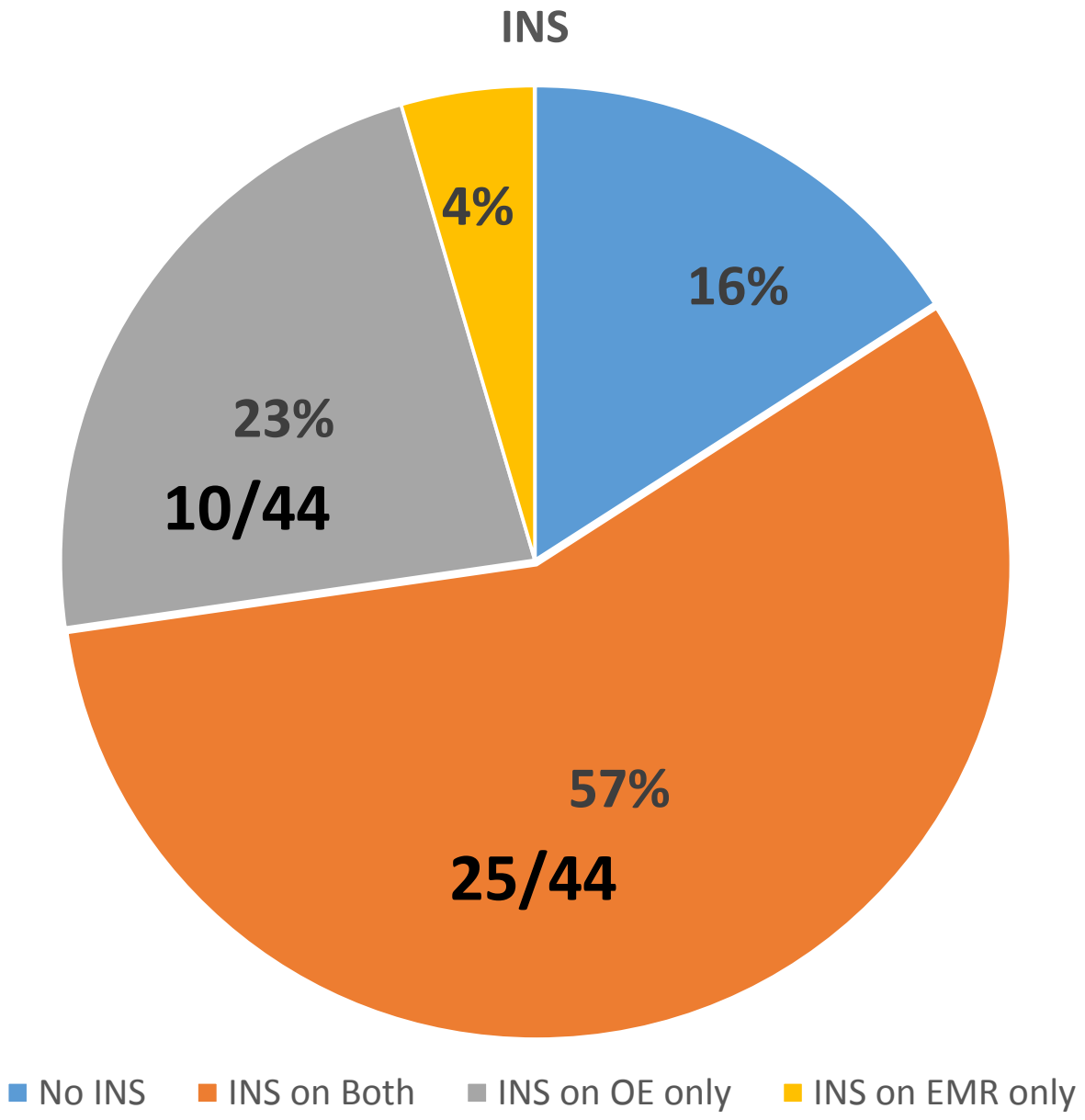
Distance convergence null ***straightens both turn & tip***

INS – OE vs EMR

- EMR and OE are significantly different in picking up INS (p=0.043)

- INS was diagnosed by Office examination BUT **not confirmed** by EMR in 10/44 cases (23%)

	EMR negative	EMR positive
OE negative	7	2
OE positive	10	25



Fusional Maldevelopment Nystagmus Syndrome (FMNS) Features that are assessed in office

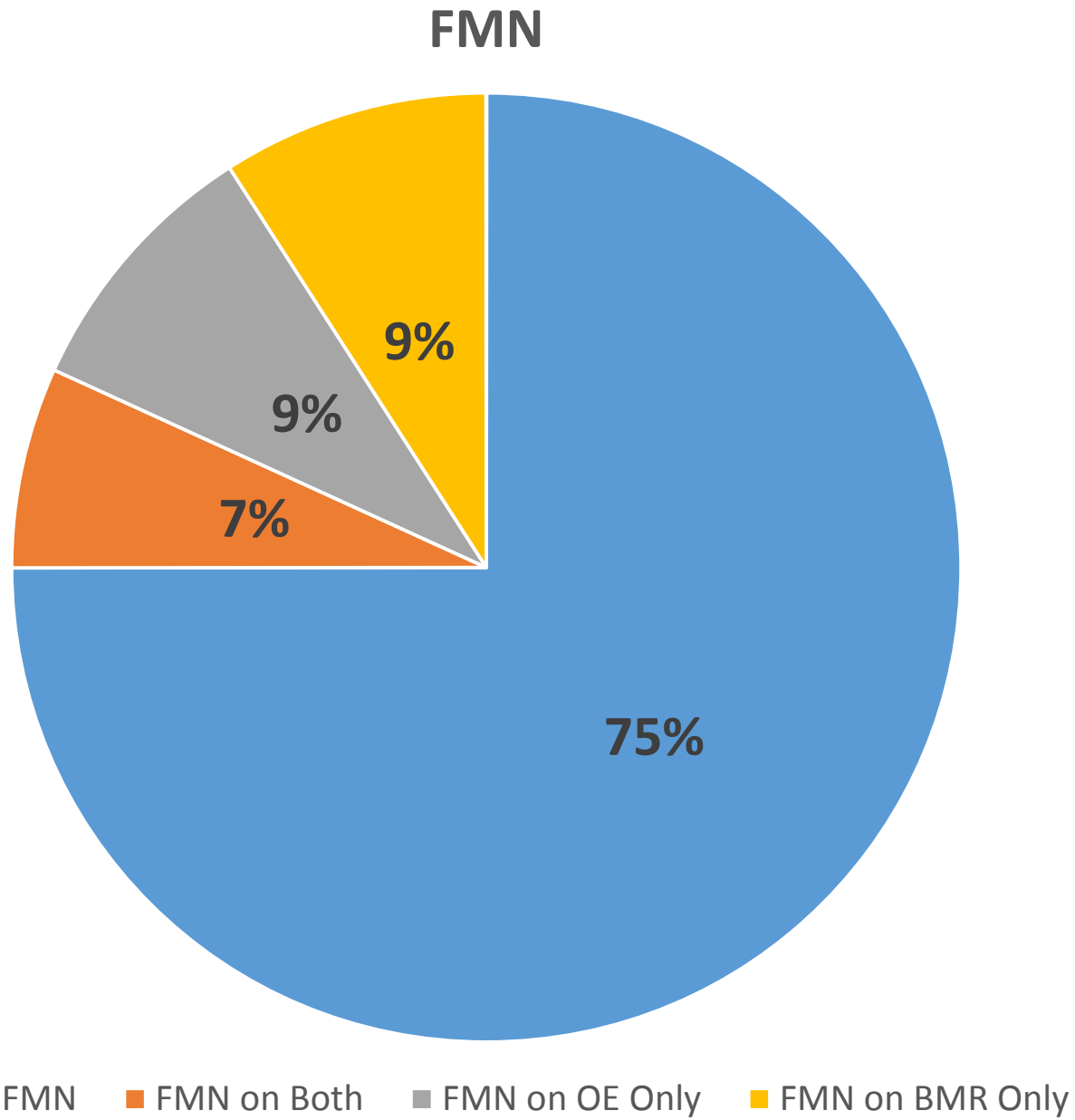
- Conjugate, horizontal, uniplanar
- Usually no associated sensory system deficits
- May dampen with exaggerated convergence (“nystagmus blockage”)
- Dissociated strabismus may be present
- Nystagmus Decreases with increased fusion (binocular function)
- Head posture associated with fixing eye in adduction

FMN – OE vs EMR

- EMR and OE were NOT different in picking up **FMN** (p=1.00)

	EMR negative	EMR positive
OE negative	33	4
OE positive	4	3

FMN = Fusion Maldevelopment Nystagmus



Aperiodic Periodic Alternating Nystagmus (APAN) clinical features

- Alternating face turns (PAN) – intervals may be regular (periodic) or irregular (aperiodic)

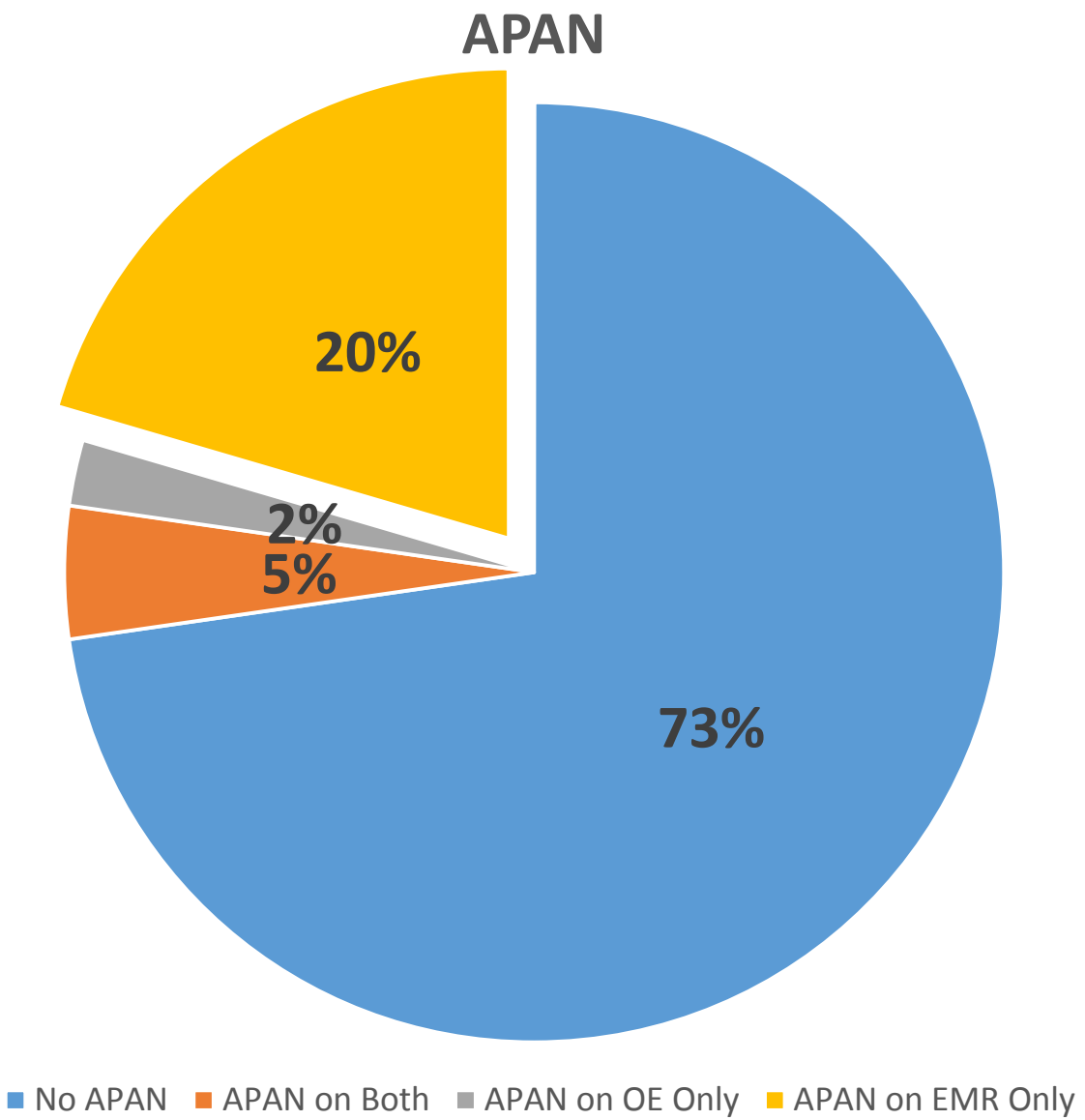
- Non-preferred eye is occluded and the preferred eye is examined with the head straight and gaze in primary position to check for alternation of nystagmus, face turns

APAN- OE vs EMR

- EMR and OE are significantly different in picking up APAN (p=0.021)

	EMR negative	EMR positive
OE negative	32	9
OE positive	1	2

APAN = Aperiodic Periodic Alternating Nystagmus



Results Summary:

- There was **disagreement** between OE and EMR for **Infantile Nystagmus (IN)** in 23% of cases.

- There is a **higher level of agreement** between OE and EMR **when there is no strabismus (p=0.038)**.

- EMR and OE were **not different** in recognising **Fusional Maldevelopment Nystagmus** (FMN) (p=1.00).

- **EMR is superior to OE** in detecting Aperiodic Periodic Alternating Nystagmus (APAN) (p=0.021), particularly in patients with sensory anomalies and those with strabismus.

Discussion and Conclusions

- When you have classic features of **INS with No Strabismus** , Eccentric Null with constant FT ± Convergence Null → **EMR is Probably not necessary and will probably not change Management**

- APAN can be easily missed in Office Examination and misdiagnosed at INS

- EMR is better than office Examination alone when there is strabismus