

Case Study of a Child with Congenital Ocular Motor Apraxia (COMA)

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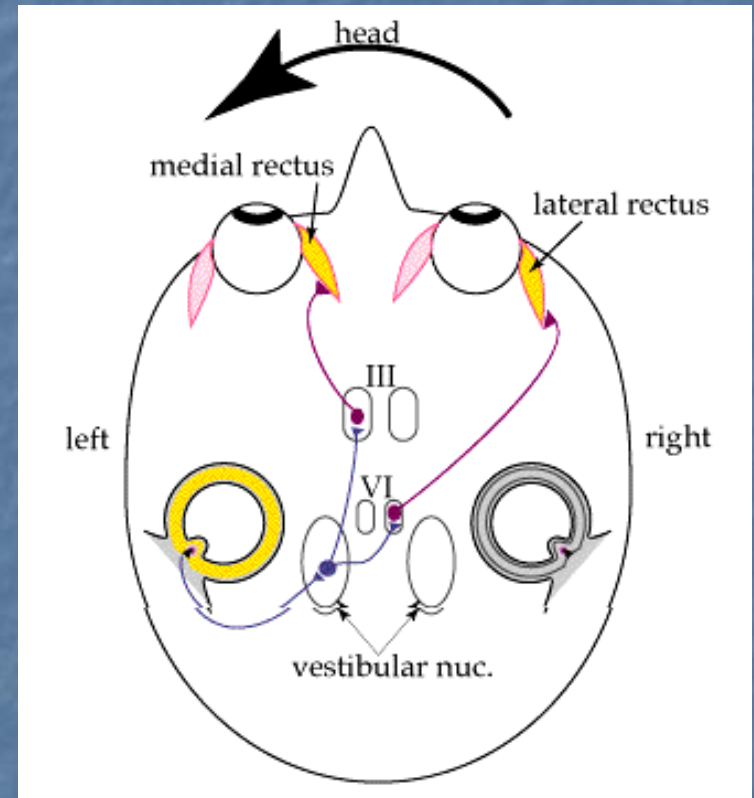
Alison

- 08/01/2009
- 8 years old
- From 2002 seen at hospital
- COMA - Congenital Ocular Motor Apraxia and high myopia



Congenital Ocular Motor Apraxia

- 1952 Professor David Logan, ophthalmologist
- Congenital Ocular Motor Apraxia (COMA)
- All vertical conjugate movements were intact
- Lateral movements were abnormal
- Characteristic head movement
- Delays in walking, slight co-ordination defects and difficulties with reading



Congenital Ocular Motor Apraxia

- 1997, Michael Sargeant, radiologist
- Voluntary horizontal saccades were impaired but random saccades, vertical eye movements and pursuit movements were normal
- MRI scans to investigate the cerebella of nineteen children with COMA
- 12/19 subjects had cerebellar abnormalities, particularly a small vermis

Congenital Ocular Motor Apraxia

Sargeant cont'd

- Feeding difficulties, speech apraxia or more generalised motor problems
- COMA head thrusts do diminish with age, when they are replaced by blinking. The speech apraxia does not improve with time.
- COMA can be autosomal dominant or recessive inheritance

Congenital Ocular Motor Apraxia

2003 Goncalves Carrasquinho

- Head thrusts appear between 4-8 months old, which was when most children were diagnosed
- Head thrusts begin with a blink to break fixation
- In adult patients the head thrust was omitted, and the change in fixation managed with a blink, although head thrusts did return when the patient was tired or distressed

Congenital Ocular Motor Apraxia

Carrasquinho cont'd

- Cases of impairment of vertical saccades reported too
- COMA can be associated with other conditions, particularly Joubert's syndrome of hypotonia, progressive ataxia, global developmental delay, sleep apnoea and hyperpnea

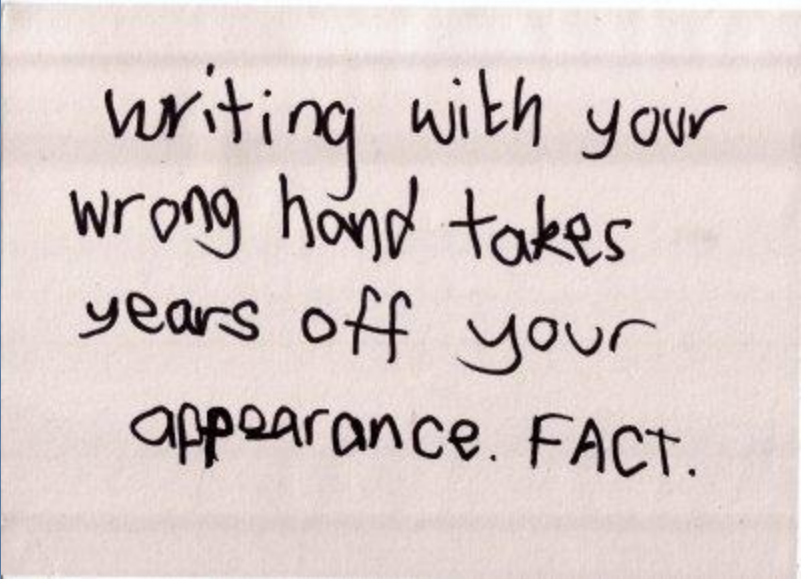
c-SIF

- COMA is a rare condition - only 50 cases reported - actual incidence higher?
- Congenital Saccade Initiation Failure (c-SIF) is a more appropriate name.

The site of the lesion is still unconfirmed, but current thinking is that it is a disorder of the frontal eye fields, sometimes in the presence of a vermian disorder of the cerebellum.

Back to Alison

- Learning difficulties at school
- Missing out words when she was reading
- Found the writing in the school books too small to read
- Getting extra help in school
- Having worksheets enlarged
- Sitting at the front of the class
- Handwriting was large and occasionally cursive
- Used guidelines when writing



writing with your
wrong hand takes
years off your
appearance. FACT.

Alison

- Little speech - yes/no, smile
- Didn't like PE or sport at all
- Couldn't swim
- Just learning to ride a bike

- Alison didn't crawl, just rolled over from about eighteen months old
- Walked at three and a half years old
- General health was good, and no meds



Alison

- R -9.75/-2.25 x 5 6/24 (20/80)
- L -14.00/-3.25 x 5 6/30 (20/100)

I found:

- R -8.00/-2.25 x 180 6/19 (20/65)
N18 amplitude 25cm
- L -15.50/-3.00 x 175 6/30+2 (20/100 +2)
N34 amplitude 20cm

Alison

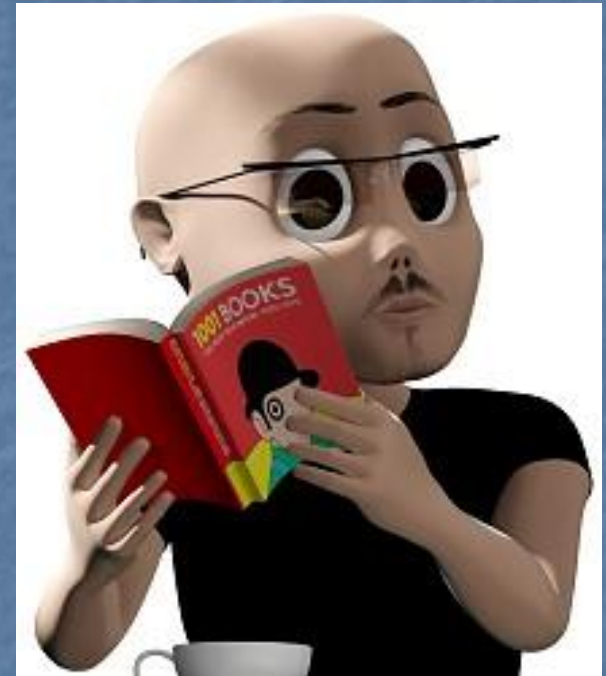
Motility - following a ball target.

- She found it very difficult to follow at all
- Looking R she showed full action but the L was slower than the R
- Looking L both eyes had restricted movements
- No action looking up and out L
- Small action on looking down and out L

Alison

Reading

- Head posture to the left
- Large HM on sweep
- Read N12 of an age 8/9 chart with good comprehension
- No deviation on cover test at dist, at near LDS?
- No convergence movements.
- Normal pupil responses
- Left suppression on Worth four dot
- No stereopsis on Randot



Maples

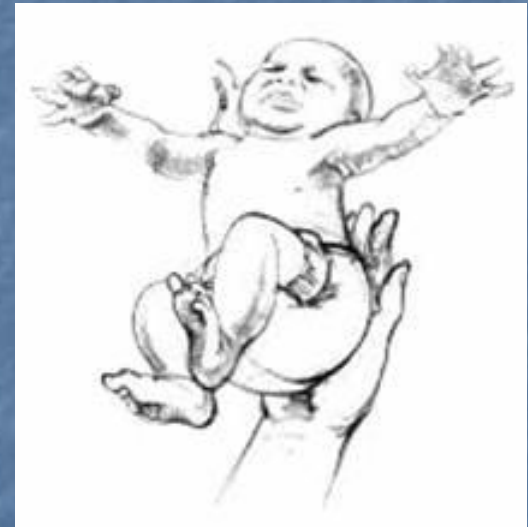
	8.1.09
Pursuit Ability	5
Accuracy	1
Head movement	1
Body movement	4
Saccades only by head movement	
Saccade Ability	5
Accuracy	1
Head movement	1
Body movement	4

Alison

Alison was very lightweight, limp, floppy, her legs looked like matchsticks and her elbows were hyper extended.

Primitive reflexes:

- TLR 2
- ATNR 2
- STNR 1
- Spinal Galant 2
- Moro 4



Alison

- Unable to crawl on her tummy
- Could crawl on all fours
- She couldn't hop on either leg, or skip
- Unable to balance in the Hot Lizard exercise

Alison had both horizontal and vertical saccadic difficulties + poor pursuit movements.

Pursuit problem may not be attributable to the COMA but more likely due to her general motor delay?

Definitely had speech apraxia.

Alison

I started by changing her glasses prescription

- TLR and Moro
- Crawl on her tummy
- Hop then skip
- Adding in ATNR, Hot Lizard and Randolph Shuffle



Alison

	8.1.09	4.3.09
■ TLR	2	0
■ ATNR	2	Slight
■ STNR	1	3
■ Spinal Galant	2	L 2
■ Moro	4	3

Alison

4/3/09

- Could crawl on her tummy, hop, skip and balance well with the Hot Lizard exercises
- Randolph Shuffle sequence unaided
- Consultant and told to wear her old glasses as mine weren't good enough!
- On the Maples test both directions on pursuits, saccades in both directions, without any head movements

Alison

Continued OVT.....

- STNR, Spinal Galant, Bear walking and Infinity Walking, improving her core stability, strength, peripheral awareness, balance and timing. Then closed eye and open eye rotations and jump fixations.
- Able to go in the garden on her own
- Beginning to do PE at school

In July Orthoptist grudgingly agreed that she did have eye movements but said that she now had a squint and nothing could be done about that.

Alison

Full reassessment on 09/12/2009.

- School work much easier
- Able to read much smaller print
- Enormous changes in her self confidence
- PE at school
- Learning to ride a bike
- Teachers had noticed the changes

Alison

Acuity 8.1.09

- R 6/19 (20/65) N18 amplitude 25cm
- L 6/30+2 (20/100 +2) N34 amplitude 20cm

- L suppression

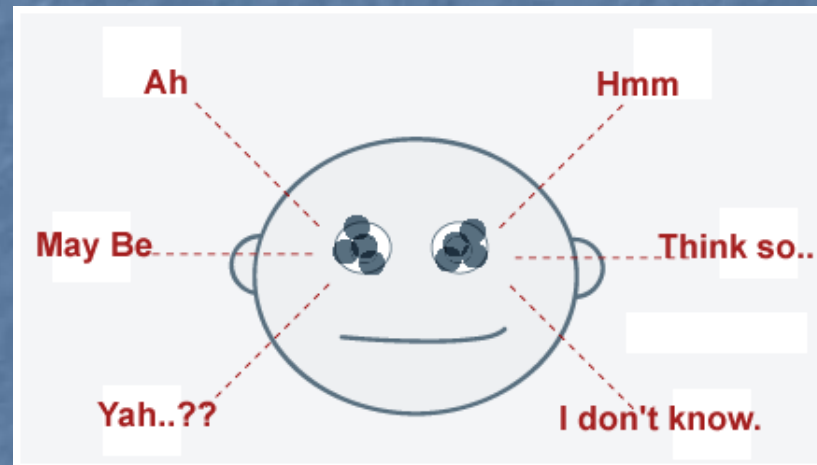
Acuity 9/12/09

- R 6/12 (20/40) N12 to 10 cm
- L 6/19 (20/65) N18 to 10cm

- L suppression

Alison

- Small XO at near on cover test, good recovery
- Convergence movements
- No stereopsis on Randot
- Motility was now full, all movements present - to the left were less smooth than to the right



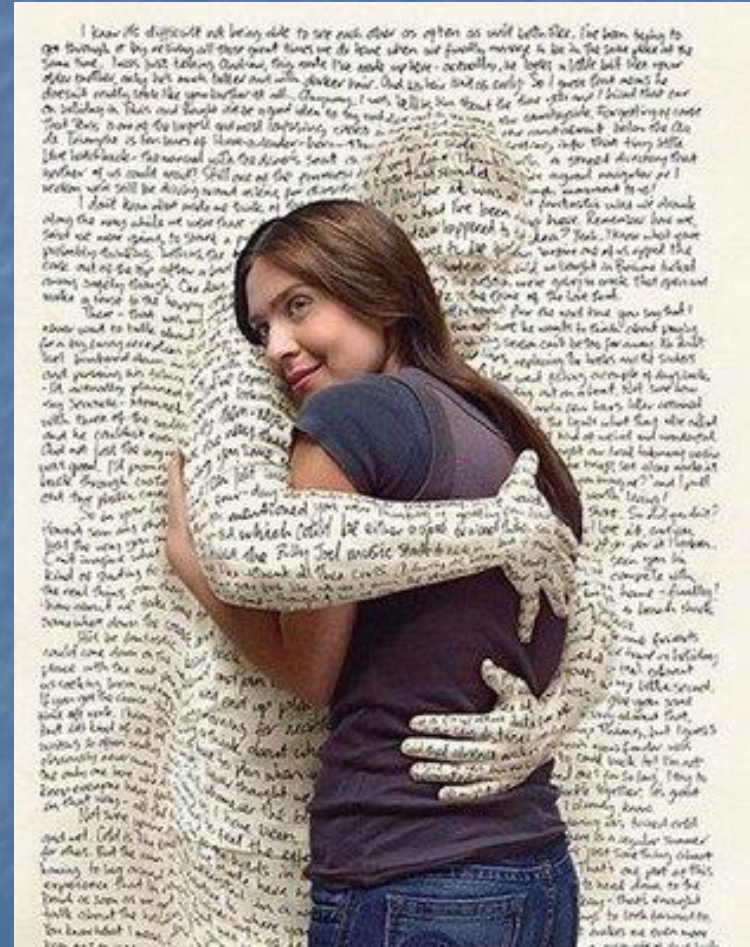
Alison

Reading 8/1/09

- Read N12 of an age 8/9 chart
- Head posture to the left
- Large HM on sweep
- Good comprehension

Reading 9/12/09

- N8 print of an age 9/10 chart
- Holding the book to the R
- Occasional HM on sweep
- Normal wd



Maples

	8.1.09	9.12.09
Pursuit Ability	5	5
Accuracy	1	1
Head movement	1	4
Body movement	4	5
Saccades only by head movement		
Saccade Ability	5	5
Accuracy	1	1
Head movement	1	4
Body movement	4	5

Alison

	8.1.09	4.3.09	9.12.09
■ TLR	2	0	0
■ ATNR	2	Slight	0
■ STNR	1	3	Slight
■ Spinal Galant	2	L 2	Slight
■ Moro	4	3	0

Alison

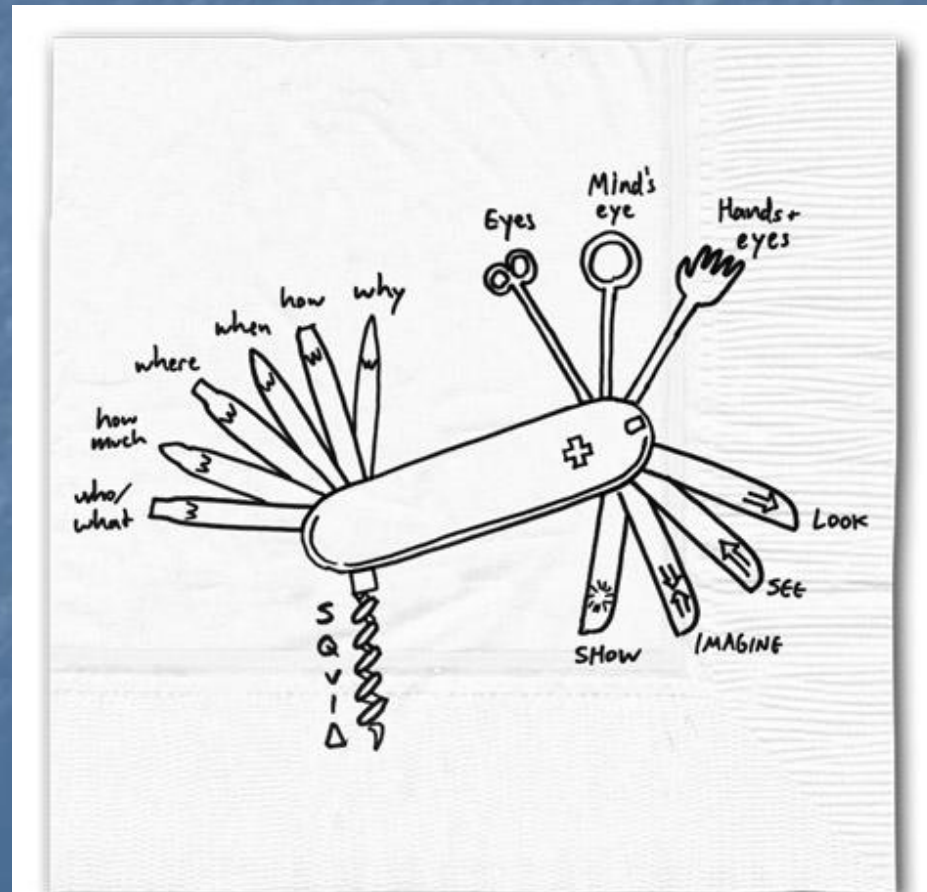
I want to:

- Recheck Rx
- Improve motor control
- Improve acuity further
- Gain binocularity
- Improve speech



Alison

'It would have got better anyway!'



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