Risk of serious intracranial pathology in children presenting with acute acquired comitant esotropia (AACE): A 20-year retrospective review

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Acute Acquired Comitant Esotropia

Characterized by:

- Sudden onset
- Manifest deviation
- Nonaccommodative

Historically, three categories:

- Type I – disruption of fusion
- Type II – physical/psychological stress with low hyperopia
- Type III- myopia

Rarely reported secondary to intracranial pathology

Purpose

To determine the risk of serious intracranial pathology in children presenting with AACE

Methods

- Retrospective chart review was performed of all children presenting to the Hospital for Sick Children with AACE who underwent neuroimaging from January 2000 to November 2020.
- Cross-checked departmental strabismus surgery database to identify additional children who presented with AACE but had neuroimaging done externally.
- Inclusion criteria:
  - Acute onset esotropia presenting within 12 months of onset
  - Comitance as defined by esotropia <5 prism diopters difference in any field of gaze
  - Neuroimaging including magnetic resonance imaging (MRI) or computerized tomography (CT)

Table 1. Demographics and Examination Findings of Children with AACE

Patient Characteristics	
Population (N)	108
Age – years (mean)	5.55
Gender	
Male	62 (57.5%)
Female	46 (42.5%)
Laterality	
Right	47 (43.5%)
Left	51 (47.2%)
Alternating	10 (9.3%)
Time to Presentation (weeks)	9
Symptoms	
Diplopia	48 (44.4%)
Headache	10 (9.3%)
Nausea and/or Vomiting	3 (2.8%)
Tinnitus	0 (0.0%)
Other Neurological Symptoms	6 (5.6%)
Examination Findings	
Amblyopia	13 (12.0%)
Size of deviation at distance (prism diopters)	31
Size of deviation at near (prism diopters)	36
Refractive Error (diopters)	1.08
Optic Nerve Edema	0
Nystagmus	1
Imaging	
Computed Tomography	16 (14.8%)
Magnetic Resonance Imaging	87 (80.6%)
Both	5 (4.6%)

Table 2. Neuroimaging Results in Children with AACE

Neuroimaging Findings	
Cerebellar medulloblastoma (case 1)	1 (0.9%)
Diffuse intrinsic pontine glioma (case 2)	1 (0.9%)
Chiari I Malformation	2 (1.8%)
Atypical, non-serious	26 (23.9%)
Normal	79 (73.5%)

Case Discussion

Case 1:

- Two-year old who presented with AACE
- No other ophthalmic or neurological findings
- Successfully treated with chemotherapy and surgical resection
- Disease free 7 years after diagnosis

Case 2:

- Eight-year-old who presented with AACE
- Complained of headaches and nausea
- Gaze-evoked nystagmus noted on exam
- Palliative radiation therapy
- Disease ultimately fatal

Conclusion

In this cohort of children with AACE, there was a small but non-trivial risk of serious intracranial pathology. When a child presents with AACE, the small risk of serious pathology must be weighted against the risks and costs of a workup. In these cases, it is advisable to either obtain neuroimaging or monitor closely for the development of concerning signs or symptoms.