Sports/activities and ALD
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15:00 Dr Gerald Raymond- Sports and ALD
Can Head Injury Influence the site of Demyelination in Adrenoleukodystrophy?

Wilkinson et al DMCN 1987, 29:797-800

It is possible that significant head trauma can activate ALD in those with ALD who were not previously presenting symptoms.
Case 1 a boy was involved in an MVA at 4 years, briefly unconscious with a left parietal fracture.

7 months later there was extensive disease involving the left hemisphere.

There are very few case studies of this.
In this instance, the blood-brain barrier is compromised.
Case 2 compound depressed skull fracture of the left posterior frontal region when accidentally hit with a golf club.

Craniotomy performed

5 years later noted to have a progressive disorder.
Very-late-onset adrenoleukodystrophy: Possible precipitation of demyelination by cerebral contusion

M. Weller, MD; W. Liedtke, MD; D. Petersen, MD; H. Opitz, MD; and M. Poremba, MD

**NEUROLOGY** 1992;42:367-370

- 57 yo man
- April 1988 left frontotemporal cerebral contusion
- Returned to work August 1988
- Deteriorated between Jan – April 1989
- Seen in December 1989, hypomanic, disinhibited, with abnormalities of gait; bronzing of skin; and alopecia
Head trauma can initiate the onset of adreno-leukodystrophy


Reported 5 cases following moderate to severe Head trauma
2 were conversions from AMN
Clinical courses were rapid
An environmental factor that is profoundly deleterious
Avoidance of potential head trauma and rapid response seemed prudent
Dramatic worsening of adult-onset X-linked adrenoleukodystrophy after head trauma

Bouquet, Floriane; Dehais, Caroline; Sanson, Marc; MD, PhD; Lubetzki, Catherine; MD, PhD; Louapre, Celine; MD, PhD

DOI: 10.1212/WNL.0000000000002173

Encephalic CT and MRI of a patient with adult-onset X-linked adrenoleukodystrophy revealed after head trauma: Figure: (A-D) Axial CT at day 0 (D0) and day 1 (D1) after a severe head trauma; red arrowheads point to the cerebral hemorrhage at the contusion site. (E, F) Axial T2-weighted FLAIR MRI and (G, H) axial T1-weighted MRI, 1 month (M1) after head trauma; red arrowheads point to sequelae of hemorrhagic lesions. No white matter demyelination was visible. (I, J) Axial T2-weighted MRI and (K, L) axial T1-weighted MRI after gadolinium injection, 5 months (M5) after head trauma, showing leukoencephalopathy predominating in the left temporal and frontal lobes. Blue arrowheads indicate lesional gadolinium enhancement. (M, N) Axial T2-weighted FLAIR MRI and (O, P) axial T1-weighted MRI after gadolinium injection, 7 months (M7) after head trauma, showing diffuse bilateral leukoencephalopathy and massive gadolinium enhancement (blue arrowheads). FLAIR = fluid-attenuated inversion recovery.
Onset of Adreno-Leukodystrophy After Medulloblastoma Therapy: Causal Connection or Coincidence?

G. Deib, A. Poretti, A. Meoded, K. J. Cohen, G. V. Raymond, M. Abromowitz, and T. A. G. M. Huisman

- Diagnosed at birth because of positive family history
- Elevated VLCFA
- MRI at age 2 normal
- 32 months presented with vomiting
- MRI demonstrated a posterior fossa tumor
- Resected
- Medulloblastoma without evidence of metastatic spread
- Treated with 5 cycles of standard chemotherapy and radiation
- MRI on completion showed no signs of tumor or ALD issues
- 14 months later – multiple metastatic lesions in the brain and spinal cord
- Chemotherapy
- MRI at 4.3 years
  - Tumor had shrunk
  - Now had WM tract involvement including splenium
  - Truncal ataxia, speech and language difficulties
Another surgical example

Presented with headaches with vomiting
Biopsied and diagnosed with MS

When seen at Minnesota for BMT
Inflammatory Demyelination

Blood-brain barrier

Capillary (in general)
- Fenestration (with intact basal membrane)
- Astrocyte
- Basal membrane
- Endothelial cell
- Intercellular space
- Mitochondria
- Tight Junctions

Capillary (Brain)
- Pericyte
- Blood
- Transport vesicles

Image: Microscopic view of a brain tissue sample showing demyelination.
Brain Injection of Lysophosphatidylcholine (C24:0) Induces Microglial Apoptosis
Definite risk factors for developing cerebral ALD

• Open head injuries
• Cerebral contusions
• Neurosurgical procedures including brain biopsy
• Severe adrenal crisis with hypotension

Risk factors for developing ALD include:
Open head injuries
Cerebral contusions
Neurosurgical procedures, including brain biopsies
Severe adrenal crisis with hypotension
Unkn0wns

- Closed head trauma without hemorrhage
- Concussions
- Spinal cord procedures

It is not clear what risk there is in closed head trauma, concussion or spinal cord procedures.
PEARLS BEFORE SWINE  

BY STEPHAN PASTIS

DON'T TELL ME YOU'RE STILL PLAYING CRICKET WITH LITTLE LEMMINGS AS BALLS.

What was that?

DON'T YOU READ THE NEWS ABOUT HEAD INJURIES IN SPORTS? HERE YOU ARE KICKING AROUND LEMMINGS WITH MAGNETS.

What are we supposed to do?

AT LEAST GIVE THEM PROTECTIVE HEADGEAR. SOMETHING STRONG THAT WILL PROTECT AGAINST CONCUSSIONS.

SANTA HATS ARE NOT PROTECTIVE.

Hey... Santa no get concussin.
Recommendations

• Allow participation in sports
  • Proper Equipment
• Close monitoring of at risk individuals who have had moderate head injury
• Combine bone marrow transplant assessment with neurosurgical interventions?

Those with an ALD diagnosis who are not presenting symptoms should be permitted to participate in sports with correct equipment and training. At risk individuals with head injuries should be closely monitored. Bone marrow transplant assessments should be combined with neurosurgical interventions.