Charles E. Niesen, MD
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Pasadena, CA
1. Typical brain injuries in child abuse

2. Anatomy and physiology of brain injury, including retinal hemorrhages

3. Medical disorders that increase risk for brain injury (differential diagnosis)

4. Differences between accidental and inflicted trauma (secrets of the trade)
ORIGINS of SHAKEN BABY SYNDROME

1962- C. Kempe “battered child syndrome”

1972- J. Caffey “whiplash shaken-baby syndrome”

Clinical, biomechanical and radiologic studies

Key features in nonaccidental head injury in infants and toddlers

Important role of angular deceleration

Hallmarks of child abuse:

1. Subdural hematoma
2. Metaphyseal (bone) fractures
3. Retinal hemorrhages
BIOMECHANICAL FORCES:

Caffey - Stressed the importance of deceleration injury

Brain = moveable mass

Repeated decelerations produce severe injury to brain and intracranial blood vessels
BIOMECHANICAL FORCES:

Recent studies- Rotational >> translational (linear) acceleration

Impact against non-yielding surface

“Shaking-impact syndrome”
BIOMECHANICAL FORCES - Meninges:
BIOMECHANICAL FORCES- Meningeal veins:

Tearing of meningeal veins →
BIOMECHANICAL FORCES- Subdural hematoma (SDH):

Small SDH

Medium SDH- mass effect
BIOMECHANICAL FORCES - Subdural hematoma (SDH):

1. Seizures
2. Motor deficits, e.g. weakness
3. Ischemic injury
4. Increased intracranial press. loss of consciousness
5. Herniation
SUBDURAL HEMATOMA- Interhemispheric fissure:

? Specific for shaken baby syndrome
Not seen in accidental head trauma in older children
METABOLIC FORCES:

Hypoxic brain injury - Lack of oxygen to diffuse or local brain areas

Direct brain injury leads to neuronal death/loss

Causes: Suffocation
Choking
Apnea (restrict chest movement)
Local changes (mass effect of subdural)

“Shaken-impact-suffocation syndrome” - never adopted

Shaking alone cannot produce constellation of injuries
METABOLIC FORCES- Hypoxic injury:

1. Suffocation
2. Choking
3. Strangulation
4. Chest compression
METABOLIC FORCES- Hypoxic injury:

Early stage: edema

Late stage: Cell death
BIOMECHANICAL and METABOLIC FORCES:

A. Infant was shaken

B. Infant hit his/her head

C. Infant was suffocated

D. Two of the above

E. All of the above

Forces that cause brain injury are multiple and additive

Severity of injury = vector, magnitude and frequency of force

Despite compound forces, the name remains the same…

Shaken baby syndrome
RETINAL HEMORRHAGES

One of the most important, if not crucial, elements in the Shaken baby syndrome triad

Etiology - Still debated

Combination of 1) increased intracranial pressure and 2) increased venous pressure

Fragility of developing retinal blood vessels

Role of angular deceleration - uncertain
RETINAL HEMORRHAGES - Mechanism:

Optic nerve/Eye - part of the brain
RETINAL HEMORRHAGIES - Mechanism:

Optic nerve sheath - extension of dura mater
RETINAL HEMORRHAGES - Mechanism:
RETINAL HEMORRHAGES- Mechanism:

Common causes of RH:

1. Crush injuries to the chest  (incr. jugular v. press.)
2. Subarachnoid hemorrhages  (incr. intracran. press.)

Traumatic head injury thought **not** to produce RH…maybe shaken baby syndrome is unique condition
Subdural hematoma + Retinal hemorrhages + Bone fracture = Child abuse

Medical dogma held for >30 years

Forces at work and fragility of developing brain = so unique that two findings together were incontrovertible

Grew into its own “subspecialty” area with new breed of pediatricians called “Child abuse specialists”

What could be easier….BUT
SHAKEN BABY SYNDROME:

Subdural hematoma + Retinal hemorrhages + Bone fracture = Child abuse

Subdural hematoma + Retinal hemorrhages + Bone fracture

Medical dogma held for >30 years

Forces at work and fragility of developing brain = so unique that two findings together were incontrovertible
SHAKEN BABY SYNDROME - New view:

Subdural hematoma + retinal hemorrhage = Many disorders

More so,

Subdural hematoma ➔ retinal hemorrhage = Many disorders
NEW PERSPECTIVE:

Subdural hematomas in infancy-

Differential diagnosis:

- Vaginal delivery
- Accidental head trauma
- Benign external hydrocephalus
- Enlarged extra-axial fluid collection
  - Normal variant
  - Prematurity
- Genetic disorders
  - Glutaric aciduria type II
  - Menkes disease
NEW PERSPECTIVE:

Retinal hemorrhages in infancy -

Differential diagnosis:
Vaginal delivery

Accidental head trauma

Chest trauma

Benign external hydrocephalus

Cardiopulomonary resusitation

Retinopathy of prematurity
NEW PERSPECTIVE - Birth trauma:

Birth trauma - Real

- One of most traumatic experiences we have
- Causes both subdural hematomas and retinal hemorrhages
- Takes 4-6 weeks for RH to disappear

<table>
<thead>
<tr>
<th></th>
<th>SDH</th>
<th>RH</th>
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<tbody>
<tr>
<td>Vaginal delivery</td>
<td>25%</td>
<td>40%</td>
</tr>
<tr>
<td>Caesarian section</td>
<td>1-3%</td>
<td>10%</td>
</tr>
</tbody>
</table>
NEW PERSPECTIVE- Accidental head trauma:

Three articles of large series that support new perspective.

Each evaluates critically the simple divide between abuse and accidental injury

1. Can short-distance falls produce severe brain injury?


   n=18 deaths from head injury on playgrounds
NEW PERSPECTIVE- Accidental head trauma:

2. What are clinical and retinal findings in children with accidental and abusive head injuries?


n= 82 (accidental: 67, abuse: 15)


n= 45 confessed abuse, 35 public accidents
NEW PERSPECTIVE- Important article:


- reviewed primary source data for all deaths involving a fall from 1988 – 1999

Results- 114 deaths, 18 due to head injury from fall

- ages: 20 mos. to 13 years,

- measured distances: 2-10 ft. (11), swings 2-6 ft. (7)

- 12/18 witnessed by noncaretaker or videotaped

- 4/6 bilateral retinal hemorrhages
NEW PERSPECTIVE- Important article:


- Full evaluation included head CT scan, serial neurologic exams, dilated ophthalmologic eye exam

Results- n= 82, (accidental: 67, abusive: 15)

<table>
<thead>
<tr>
<th></th>
<th>Abuse</th>
<th>Accident</th>
</tr>
</thead>
<tbody>
<tr>
<td>RH</td>
<td>60%</td>
<td>10%</td>
</tr>
<tr>
<td>Unilateral RH</td>
<td>20%</td>
<td>9%</td>
</tr>
<tr>
<td>Bilateral RH</td>
<td>40%</td>
<td>1%</td>
</tr>
<tr>
<td>Pre-RH</td>
<td>30%</td>
<td>0</td>
</tr>
<tr>
<td>Single RH</td>
<td>0</td>
<td>4%</td>
</tr>
<tr>
<td>Extends to periphery</td>
<td>27%</td>
<td>0</td>
</tr>
</tbody>
</table>
NEW PERSPECTIVE- Important article:

Blechtel et al. (2004)-

<table>
<thead>
<tr>
<th>Condition</th>
<th>Abuse</th>
<th>Accident</th>
</tr>
</thead>
<tbody>
<tr>
<td>Seizures</td>
<td>53%</td>
<td>6%</td>
</tr>
<tr>
<td>Altered consciousness</td>
<td>53%</td>
<td>10%</td>
</tr>
<tr>
<td>Scalp hematoma</td>
<td>7%</td>
<td>51%</td>
</tr>
</tbody>
</table>
CONCLUSIONS:

1. Minor head trauma can cause significant brain injury.

2. Accidental head trauma can cause retinal hemorrhages.

3. Accidental head trauma tends to produce unilateral RH.

4. Clinical findings are important in distinguishing abuse from accidental trauma.
NEW PERSPECTIVE:

Subdural hematomas in infancy-
Differential diagnosis:
  Vaginal delivery
  Accidental head trauma

Benign external hydrocephalus

Enlarged extra-axial fluid collection
  Normal variant
  Prematurity

Genetic disorders
  Glutaric aciduria type II
  Menkes disease
**BENIGN EXTERNAL HYDROCEPHALUS:**

**Definition:** Rapid head growth in 1\textsuperscript{st} year of life demonstrated on CT scan by enlarged subarachnoid space

**Pathophysiology:** Cause of rapid growth is uncertain
- No evidence of increasing pressure
- Skull growth >> brain growth

**Genetics:** Associated with one or more parents with macrocephaly
- No gene or gene locus identified

**Symptoms:** Enlarging head circumference
- Prominent or broad forehead
- No signs of increased intracranial pressure, e.g. vomiting, lethargy, seizures, bulging fontanelle
- No loss of developmental milestones
BENIGN EXTERNAL HYDROCEPHALUS:

Clinical course:
No seizures
Minor trauma can produce subdural hematomas
Increased subarachnoid space stretches meningeal veins almost to breaking point
Can cause “subdural hematomas of different ages”
BENIGN EXTERNAL HYDROCEPHALUS:

15 months old male- tripped and fell at playground

- hit head on hard soil
- witnessed by grandmother
- no loss of consciousness
- later, irritable and vomiting

At ER, head CT scan done- admitted for suspected child abuse
BENIGN EXTERNAL HYDROCEPHALUS:

Symmetric extra-axial fluid collection

Right chronic subdural
BENIGN EXTERNAL HYDROCEPHALUS:

“Subdural hematomas of different ages”
BENIGN EXTERNAL HYDROCEPHALUS:

Also, retinal hemorrhages found, R > L

Child removed from the home . . . . BUT

Three important facts were overlooked:

1. Rapid head circumference at 2-6 months of age

2. Dad’s head circumference >98%

3. History of forceful rocking behavior, when playful or angry
BENIGN EXTERNAL HYDROCEPHALUS:

- 2 to 6 months: rapid head growth
- 6 months: HC >98% (macrocephalic)
BENIGN EXTERNAL HYDROCEPHALUS:

Consider:

1. Head growth before onset of “abuse”
2. Both dad and son are macrocephalic.
3. He has inherited condition called “Familial Megalencephaly”
4. Alternative explanation for shaken baby syndrome called “shake it baby” syndrome
BENIGN EXTERNAL HYDROCEPHALUS:

Consider:
5. CT scan- Enlarged, symmetric extra-axial fluid collection
   - Known as “Benign external hydrocephalus”

6. More room for veins to be shaken and torn

7. Retinal hemorrhages- History of recurrent bleeds
   - this raises intracranial pressure
   - reaches “tipping point” at playground
   - one final “blow” that ruptures vessels
SUBDURAL HEMATOMA - Distinguishing subdural from subarachnoid space
SUBDURAL HEMATOMA - Distinguishing subdural from subarachnoid space

Sagittal view

Axial view
SUBDURAL HEMATOMA - Distinguishing subdural from subarachnoid space

Large, bilateral subdurals (nonaccidental)

Large, bilateral subarachnoid (accidental)
BENIGN EXTERNAL HYDROCEPHALUS (BEH):

What’s in a name?

Is it hydrocephalus? NO. No increased pressure or enlarged ventricles.

Is it benign? NO. Increased risk for hematomas. Children removed from homes for suspected abuse.

Is it external? POSSIBLY. Doesn’t emphasize true nature of problem.

Incidence: macrocephaly- 2% of children suggests ~ 0.5-1% have BEH, 1/50-100
Be on the look out!
NEW PERSPECTIVE- Important article:

Vinchon M. et al. (2010)- Prospective study of <2 year old children admitted for head trauma

- Evaluation included head CT scan, serial neurologic exams, ophthalmologic exam from 2001 to 2009

Results- 419 cases of head injury, 45/124 abusive and 39/288 accidental were corroborated, i.e. abuser confessed, accident in public place witnessed

n= 84 patients
NEW PERSPECTIVE - Important article:

<table>
<thead>
<tr>
<th></th>
<th>Inflicted</th>
<th>Accident</th>
<th>p</th>
</tr>
</thead>
<tbody>
<tr>
<td>N</td>
<td>45</td>
<td>39</td>
<td></td>
</tr>
<tr>
<td>M/F</td>
<td>29/16 (1.81)</td>
<td>23/16 (1.44)</td>
<td>0.24</td>
</tr>
<tr>
<td>Perinatal illness</td>
<td>22 (48.9%)</td>
<td>11 (28.2%)</td>
<td>0.055</td>
</tr>
<tr>
<td>Socio-psy</td>
<td>20 (44.4%)</td>
<td>2 (5.1%)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Stepparent</td>
<td>5</td>
<td>1</td>
<td>NS</td>
</tr>
<tr>
<td><strong>Age (months)</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Mean</td>
<td>3.8</td>
<td>8.1</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Median</td>
<td>3.2</td>
<td>5.5</td>
<td></td>
</tr>
<tr>
<td>Extremes</td>
<td>0.8–18.3</td>
<td>0–23.9</td>
<td></td>
</tr>
<tr>
<td>Delay to referral (hours)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Mean</td>
<td>57.0</td>
<td>21.2</td>
<td>0.09</td>
</tr>
<tr>
<td>Median</td>
<td>12.0</td>
<td>3.5</td>
<td></td>
</tr>
<tr>
<td>Extremes</td>
<td>0–646</td>
<td>0–396</td>
<td></td>
</tr>
<tr>
<td>Delay to CT (days)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Mean</td>
<td>2.3</td>
<td>0.5</td>
<td>0.026</td>
</tr>
<tr>
<td>Median</td>
<td>0.8</td>
<td>0.1</td>
<td></td>
</tr>
<tr>
<td>Extremes</td>
<td>0–27</td>
<td>0–3.7</td>
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NEW PERSPECTIVE- Important article:

<table>
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<tr>
<th>Clinical features</th>
<th>Abused</th>
<th>Accidental</th>
<th>p-value</th>
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<tbody>
<tr>
<td>Seizures</td>
<td>31 (68.9%)</td>
<td>5 (12.8%)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Somnolence</td>
<td>18 (60.0%)</td>
<td>12 (30.8%)</td>
<td>0.38</td>
</tr>
<tr>
<td>Coma</td>
<td>20 (44.4%)</td>
<td>9 (23.1%)</td>
<td>0.04</td>
</tr>
<tr>
<td>Deficit</td>
<td>20 (44.4%)</td>
<td>8 (20.5%)</td>
<td>0.02</td>
</tr>
<tr>
<td>Raised intracranial pressure</td>
<td>30 (66.7%)</td>
<td>9 (23.1%)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Swelling</td>
<td>1 (2.2%)</td>
<td>30 (76.9%)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Impact on head</td>
<td>17 (37.8%)</td>
<td>34 (87.2%)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Other impact</td>
<td>11 (24.4%)</td>
<td>6 (15.4%)</td>
<td>0.30</td>
</tr>
<tr>
<td>Peripheral fracture</td>
<td>12 (26.7%)</td>
<td>2 (5.1%)</td>
<td>0.008</td>
</tr>
<tr>
<td>Transfusion</td>
<td>10 (22.2%)</td>
<td>10 (25.6%)</td>
<td>0.71</td>
</tr>
<tr>
<td>Life threat</td>
<td>3 (6.7%)</td>
<td>8 (20.5%)</td>
<td>0.060</td>
</tr>
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</table>
### Table 2 Radiological and ophthalmological findings in the IHI and AT groups

<table>
<thead>
<tr>
<th>CT findings</th>
<th>Inflicted</th>
<th>Accident</th>
<th>p</th>
</tr>
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<tbody>
<tr>
<td>Subdural collection</td>
<td>37 (82.2%)</td>
<td>17 (43.6%)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Mixed density image</td>
<td>39 (90.7%)</td>
<td>17 (53.1%)</td>
<td>&lt;0.001</td>
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<tr>
<td>Brain ischemia</td>
<td>12 (26.7%)</td>
<td>1 (3.2%)</td>
<td>0.0023</td>
</tr>
<tr>
<td>Extradural</td>
<td>1 (2.4%)</td>
<td>5 (13.9%)</td>
<td>NS</td>
</tr>
<tr>
<td>Contusion</td>
<td>7 (15.6%)</td>
<td>10 (25.6%)</td>
<td>0.25</td>
</tr>
<tr>
<td>Fracture</td>
<td>5 (11.4%)</td>
<td>26 (66.7%)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Total impact</td>
<td>10 (22.2%)</td>
<td>26 (66.7%)</td>
<td>&lt;0.001</td>
</tr>
</tbody>
</table>

**Retinal hemorrhage**

<table>
<thead>
<tr>
<th></th>
<th>Inflicted</th>
<th>Accident</th>
<th>p</th>
</tr>
</thead>
<tbody>
<tr>
<td>None</td>
<td>7 (15.9%)</td>
<td>29 (82.9%)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Mild</td>
<td>3 (6.8%)</td>
<td>5 (14.3%)</td>
<td>NS</td>
</tr>
<tr>
<td>Moderate</td>
<td>9 (20.5%)</td>
<td>0</td>
<td>NS</td>
</tr>
<tr>
<td>Severe</td>
<td>25 (56.8%)</td>
<td>1 (impact)</td>
<td>&lt;0.001</td>
</tr>
</tbody>
</table>
GENETIC DISORDERS:

Glutaric aciduria type II - rare cause of severe mental retardation
- Caused by defect in fatty oxidation
- **Incidence**: 1/20-50,000
- **Symptoms**: In infant, hypotonia, vomiting and hypoglycemia
- **Imaging**: Macrocephaly, increased extra-axial fluid spaces
  Increased signal change in basal ganglia
- **Diagnosis**: elevated urinary glutaric and hydroxyglutaric acid
- **Treatment**: Riboflavin and carnitine help

Menkes disease – rare cause of severe mental retardation
- Caused by defect in copper metabolism
- **Incidence**: 1/30-50,000
- **Symptoms**: developmental delay, rust-colored, kinky hair
- **Imaging**: Brain atrophy, increased extra-axial fluid spaces
- **Diagnosis**: Elevated copper levels
- **Treatment**: None
HOW GOOD IS THE EVIDENCE?

Impossible to perform randomized, doubled-blinded control trial

Two prospective case-control studies (n=84, another n=82)

Rare and common medical disorders that increase risk of minor trauma

No animal models

...HOWEVER
HOW GOOD IS THE EVIDENCE?

Simple rules on deciding the cause of head trauma in children no longer apply.

Retinal hemorrhages occur in accidental head trauma.

Pre-existing medical conditions need to be sought.

Abused children are sicker at presentation with seizures and depressed level of consciousness.

Accidental trauma shows soft tissue injury from impact.
HOW GOOD IS THE EVIDENCE?

**Medicine** is pulled into the future, kicking and screaming

**Medicine** is conservative, stubborn and skeptical

It needs to be told things over and over again

**Attorneys and the courts** have no dogma to defend

The **evidence can be weighed**, an algorithm can not apply

Vinchon et al. (2010)- want to establish a new triad:

SDH + severe RH + lack of impact = Child abuse

Yet, no mention of preexisting medical conditions, like BEH