Shaken Baby Syndrome: The Triad and Importance of Retinal Hemorrhages

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“…shaken baby syndrome is now a well characterized clinical and pathological entity with diagnostic features in severe cases virtually unique to this type of injury”

Syndrome

• A Group of Signs and Symptoms that Collectively Indicate a Disease or Disorder

Triad

• A Group of Three
Triad of SBS

1. Subdural Hemorrhage
2. Retinal Hemorrhage
3. ???????
   - Long Bone Fractures
   - Subarachnoid hemorrhage
   - Diffuse Axonal Injury
   - Encephalopathy
Donohoe M, Am J Forensic Med Pathol 2003;24: 239–242

• Conclusion

• “the data available in the medical literature by the end of 1998 were inadequate to support any standard case definitions, or any standards for diagnostic assessment.”
Retinal Hemorrhages
Anatomy of the Eye

Retinal Hemorrhages

• “When extensive retinal hemorrhage accompanied by perimacular folds and schisis cavities is found in association with intracranial hemorrhage or other evidence of trauma to the brain in an infant, shaking injury can be diagnosed with confidence regardless of other circumstances.”

  American Academy of Ophthalmology
Retinal Hemorrhages

- The overwhelming body of literature supports a conclusion that severe hemorrhagic retinopathy in otherwise previously well children without obvious history to the contrary (eg, fatal head crush) suggests that the child has been submitted to abusive repetitive acceleration-deceleration trauma with or without head impact.

- Levin AV, Retinal Hemorrhage in Abusive Head Trauma. *Pediatrics* 2010; 126(5):1-10
Causes of Retinal Hemorrhages

- Sudden Increase in Intracranial Pressure
- Bleeding Disorders
- Retinal Vein Thrombosis
- High Blood Pressure
- Severe Anemia
- Vaginal Delivery
- Trauma
Terson Syndrome

• Terson A. De l’hémorrhagie dans le corps vitré au cours de l’hémorrhagie cérébrale. *Le Clinique Ophtalmologique* 1900;6(22):309-312.

  - Describes the association between intracranial and intraocular hemorrhage

  - Published in 1900
RH – Increased ICP


• Caused retinal hemorrhage in monkeys by inflating balloons in their skulls
RH – Increased ICP

- Muller PJ, Deck JH. Intraocular and optic nerve sheath hemorrhage in cases of sudden intracranial hypertension. *J Neurosurg* 1974

  - Postmortem examination of eyes from 23 patients with sudden increased intracranial hemorrhage, and 24 patients with no evidence of increased intracranial pressure
    - Severe cerebral trauma (4)
    - Massive spontaneous intracerebral hemorrhage (8)
    - Ruptured berry aneurysm (9)
    - Internal carotid occlusion resulting in massive postinfarction cerebral swelling without hemorrhage (2)
Intraocular and optic nerve sheath hemorrhage in cases of sudden intracranial hypertension

• Results
  ➢ 87% of the eyes had optic nerve sheath hemorrhages
  ➢ 37% of the eyes had intraocular hemorrhages

• Conclusion
  ➢ “…on the basis of gross and histological examination of 46 eyes from cases of sudden intracranial hypertension, as well as a cadaver cavernous sinus effusion study and review of the venous drainage of the eye and orbit, conclude that intraocular and optic nerve sheath hemorrhages result from the transmission of intracranial pressure into the optic nerve sheath through the subarachnoid communication in the optic canal and not via vascular routes.”
• Tongue AC. The ophthalmologist's role in diagnosing child abuse. *Ophthalmology* 1991 (editorial)

>“Until it is unequivocally proven that retinal folds are secondary to dynamic vitreous traction and shaking and not some other factors, it is imperative that we not equate retinal folds with child abuse, just as we cannot equate the presence of retinal hemorrhage with child abuse.”
RH – Vitreous Traction?

- Emerson MV, Jakobs E, Green WR. Ocular autopsy and histopathologic features of child abuse. Ophthalmology. 2007
  - Postmortem examination of eyes from 118 cases of “suspected or admitted child abuse,” 7 of which admitted to shaking
  - Retinal hemorrhages were present in 41.1% of the eyes and 44.9% of the cases
  - “We propose that retinal hemorrhage in shaking abuse is caused by venous stasis and leakage from retinal vessels, which, if prolonged, can lead to circumferential macular and peripheral retinal folds with blood- and protein-filled schisis cavities.”
Retinal Hemorrhages

- Lantz PE, Sinal SH, Stanton CA, and Weaver RG. Perimacular retinal folds from childhood head trauma, BMJ 2004
  - Case Report and Review of Literature on Perimacular Retinal Folds
  - 14 month old child fatally injured when a television fell on his head
  - Bilateral dot and blot intraretinal
  - Ophthalmologic examination revealed bilateral dot and blot intraretinal hemorrhages, preretinal hemorrhages, and perimacular retinal folds
  - Autopsy confirmed bilateral retinal hemorrhages and perimacular retinal folds
Perimacular Retinal Folds from Childhood Head Trauma

- Review identified 42 articles and book chapters
- Concluded
  - “Statements in the medical literature that perimacular retinal folds are diagnostic of shaken baby syndrome are not supported by objective scientific evidence.”
  - “Noncomparative observational reports and unsystematic narrative review articles contain insufficient evidence to provide unbiased support for or against diagnostic specificity, and inferences about associations, causal or otherwise, cannot be determined.”
RH – Animal Experiment

  - Produced non-impact head injury in piglets and examiner their eyes and brains
  - No retinal hemorrhages were identified

- Two kittens and one rabbit were shaken to death by a dog and their eyes were examined
- “No evidence of vitreous haemorrhage, retinal detachment, or retinoschisis.”
- “No retinal or optic nerve sheath haemorrhage was found.”
- No mention of the cause of death
Severe RH = Shaking?

• How large of a study is necessary to prove the hypothesis that severe RH in children = Shaking is false?
  ➢ ONE

FIGURE 3. Multiple retinal hemorrhages extended to the ora serrata. The entire optic nerve sheath was darkened by marked hemorrhage.
Retinal and Optic Nerve Sheath Hemorrhages are Not Pathognomonic of Abusive Pediatric Head Injury

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Presented at the Annual Conference of the American Academy of Forensic Sciences in 2010
RESULTS

• 137 cases met age criteria between 2003 and 2007 ($N_{COMPLETE}$)

• 10 cases were excluded
  • 8 cases were signed out without autopsy
    • Known lethal natural disease
  • 2 cases did not have ‘complete’ case files

• 127 cases (92.7%) were included in the study ($N_{TOTAL}$)
RESULTS

• Manner of death certification of $N_{TOTAL}$:
  • Natural – 43 (33%)
  • Accident – 60 (47%)
  • Homicide – 12 (10%)
  • Undetermined - 12 (10%)
RESULTS

- 19 cases had eye pathology
- Eye pathology included
  - Retinal hemorrhages only – 7 (37%)
  - Peri-optic nerve sheath hemorrhage only – 4 (21%)
  - Retinal and optic nerve sheath hemorrhages – 6 (32%)
  - Retinal and perioptic nerve sheath hemorrhages plus retinoschisis – 2 (10%)
RESULTS

• COD certification for the 19 cases with abnormal eye findings
  • Blunt head trauma / blunt trauma – 12 (63%)
  • ‘Asphyxia’ – 3 (16%)
  • Drowning – 1 (5%)
  • Pneumonia – 1 (5%)
  • Seizure disorder – 1 (5%)
  • Gunshot wound of head – 1 (5%)
RESULTS

• MOD certification for the 19 cases with abnormal eye findings
  • Natural – 2 (11%)
  • Accident – 8 (42%)
  • Homicide – 9 (47%)
Conclusions

- Retinal and perioptic nerve sheath hemorrhages are not pathognomonic of pediatric inflicted head trauma.
- Retinal and perioptic nerve sheath hemorrhages are highly associated with:
  - Restitution of a perfusing pressure following resuscitation
  - Reactive coagulopathy
  - Brain swelling
  - Blunt head trauma, especially subarachnoid hemorrhage
Conclusions

• The presence and/or absence of retinal and/or perioptic nerve sheath hemorrhages does not allow investigators to differentiate between inflicted and non-inflicted trauma, nor does it make one diagnosis more likely than the other.

• If the results of a ‘test’ cannot be interpreted, then the test should not be ordered. Thus, routine eye removal should be discouraged, even in cases of alleged inflicted head trauma.
Extensive Hemorrhagic Retinopathy, Perimacular Fold, Retinoschisis, and Retinal Hemorrhage Progression Associated with a Fatal, Spontaneous, Non-traumatic Intracranial Hemorrhage in an Infant

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Oral Presentation: 65th Annual Meeting of the American Academy of Forensic Sciences; Washington, DC February 22, 2013 @ 10:30 AM (G82)
Clinical History

• 2 month old female infant with no significant medical history
• Wakes up from a nap and is fussy
• Suddenly becomes unresponsive while trying to be soothed
• CT scan reveals a large subarachnoid hemorrhage thought secondary to an aneurysm, arteriovenous malformation, or tumor
Clinical History

• Due to the diffuse nature of the blood non-accidental trauma could not be ruled out
• Osseous survey was interpreted with no evidence of acute or healing fractures
• Review of chest radiographs and osseous survey revealed healing fractures of anterior left 7th & 8th ribs (> 2 weeks and more likely 4-6 weeks old)
Timeline

- 5/24/12 @ 1912: 911 call received
- 5/24/12 @ 1920: EMS arrives at scene; family performing 2-finger CPR and mouth-to-mouth rescue breaths; EMS assessment: heart rate of 169/minute but apneic; bag valve mask ventilation; anterior fontanelle rigid, non-soft
- 5/24/12 @ 2017: Arrival in ED; intubated & mechanically ventilated
- 5/24/12 @ 2115: Cranial computed tomography
- 5/25/12 @ 0411: Osseous survey
- 5/25/12 @ 1134 PICU attending/CA consult: clinical presentation not c/w child abuse; child abuse protocol lifted but ophthalmology consult not cancelled
- 5/25/12 @ 1400: Clinical brain death examination (#1)
- 5/25/12 @ 1648: Cerebral blood flow study
- 5/25/12 @ 1740: Ophthalmology consult w/ RetCam images
- 5/25/12 @ 1800: Clinical brain death examination (#2)
- 5/25/12 @ 1824: Pronounced dead
- 5/25/12: Organ donation
- 5/26/12 @ 2345: PMIO
- 5/28/12: Autopsy
Autopsy Findings

• Subdural and Subarachnoid Hemorrhage
• Subdural Hemorrhage of Spinal Cord
• Bilateral Optic Nerve Sheath Hemorrhage
• Bilateral Retinal Hemorrhages with Circinate Retinal Folds in the Right
• Ruptured Arteriovenous Malformation of Choroid Plexus
Retinal Hemorrhage Progression with Circinate Retinal Folds (OD)

RetCam images 5/25/12 @ 1740.

PM ocular images
Shaken Baby Syndrome

Is a hypothesis accepted as truth in the medical, legal and social literature. The theory is reiterated again and again without any attempt at scientific verification, and the signs and symptoms are stated to be pathognomonic for abuse or intentional injury.
Retinal Hemorrhages

• The most often used finding to verify abusive head injury, but in reality are meaningless