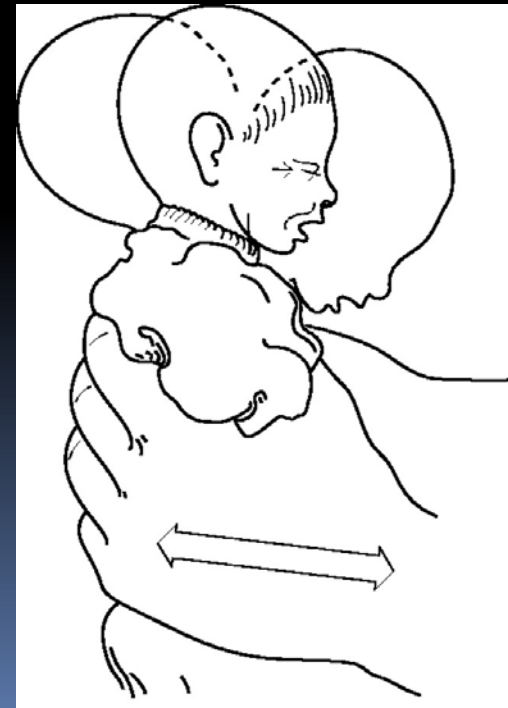


Charles E. Niesen, MD
AMS Neurology, Inc.
Pasadena, CA



NEUROLOGY 101:

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:

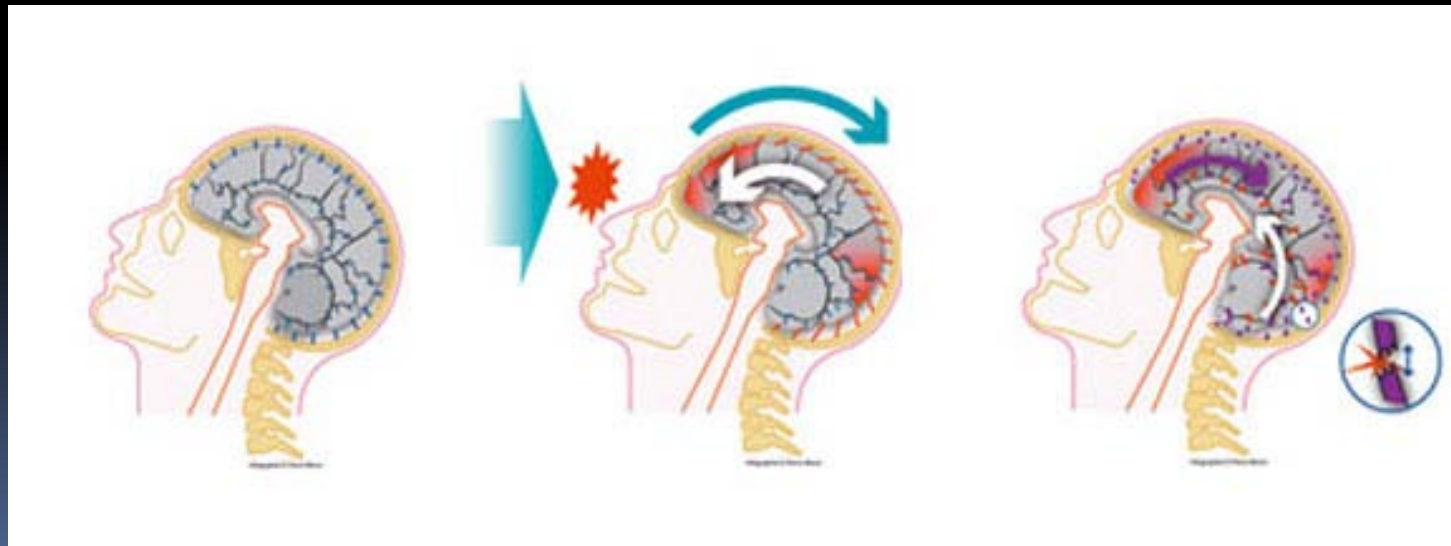
1. Subdural hematoma
2. Metaphyseal (bone) fractures
3. Retinal hemorrhages

BIOMECHANICAL FORCES:

Caffey- Stressed the importance of deceleration injury

Brain = moveable mass

Repeated decelerations produce severe injury to brain and intracranial blood vessels

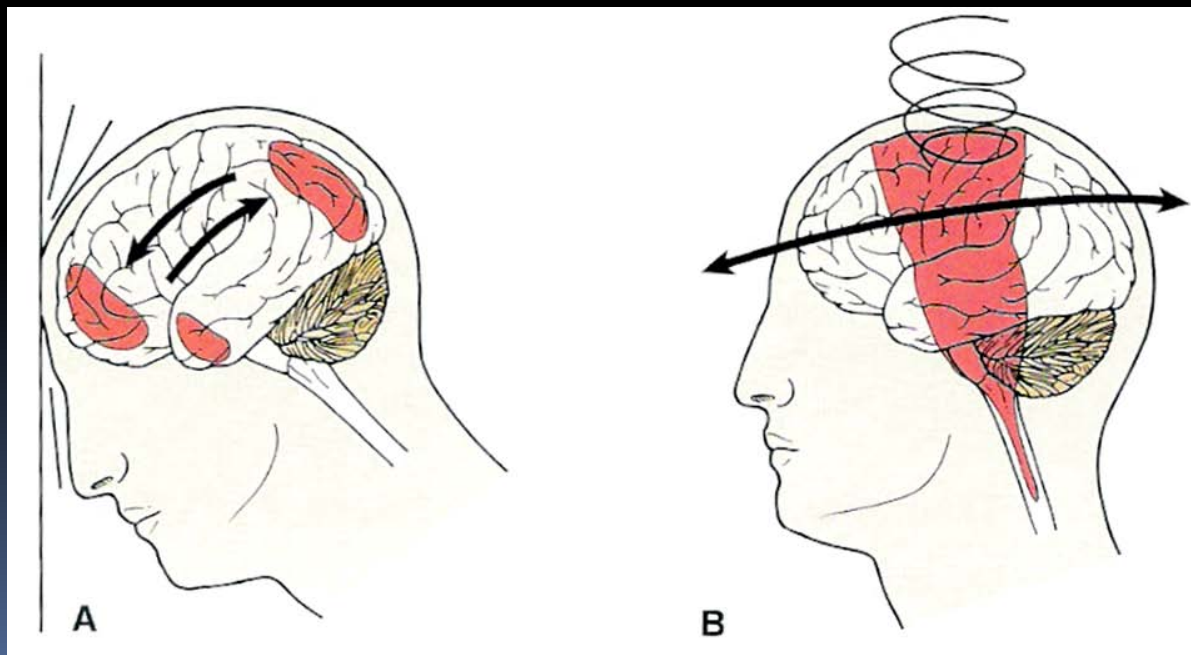


BIOMECHANICAL FORCES:

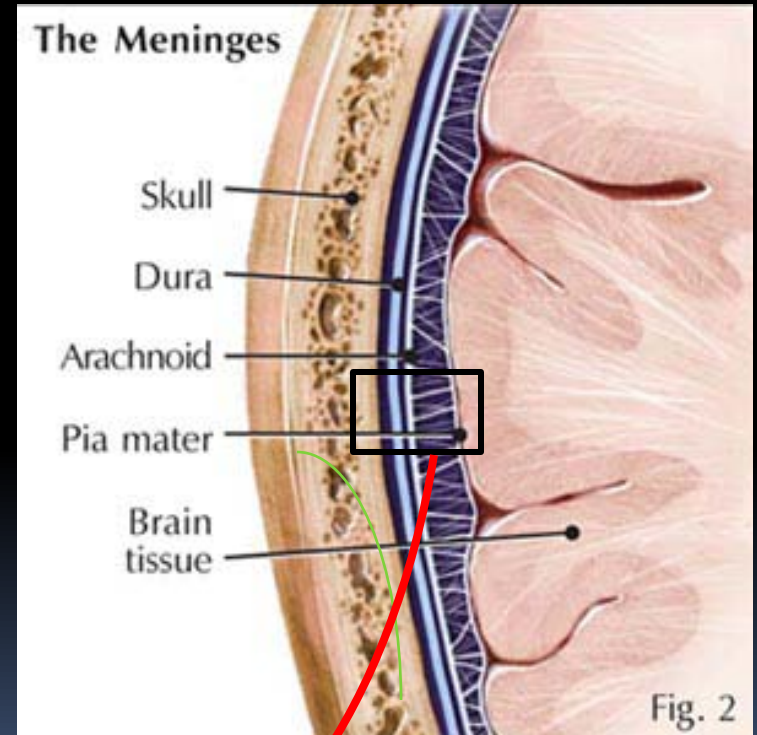
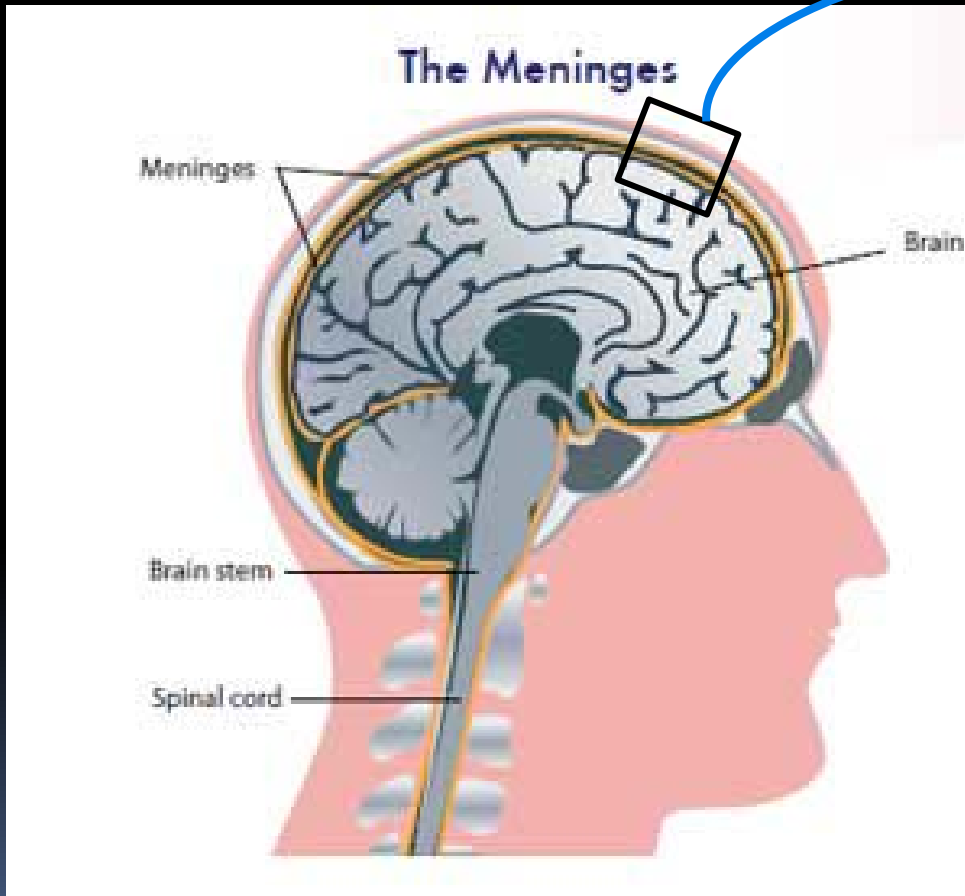
Recent studies- Rotational \gg translational (linear) acceleration

Impact against non-yielding surface

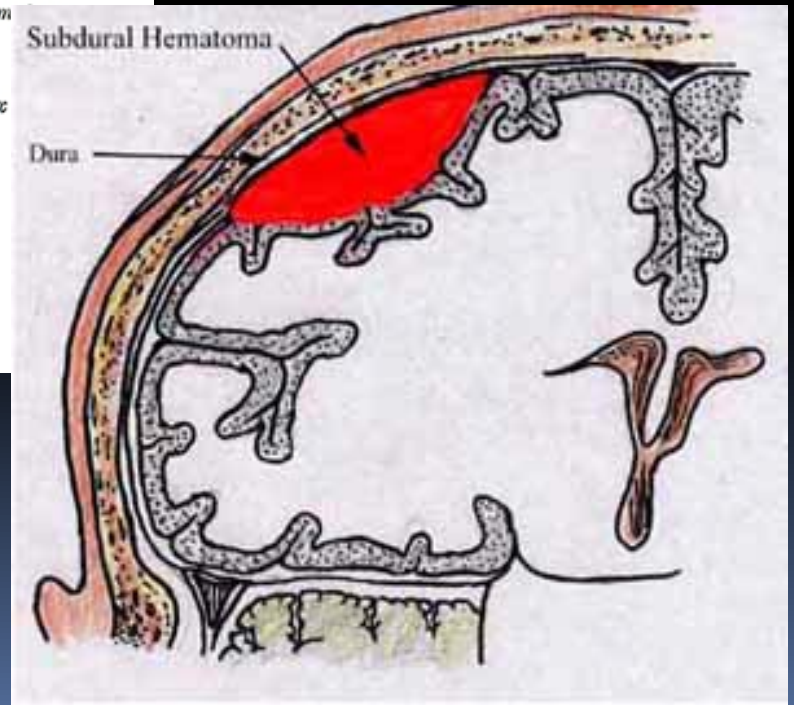
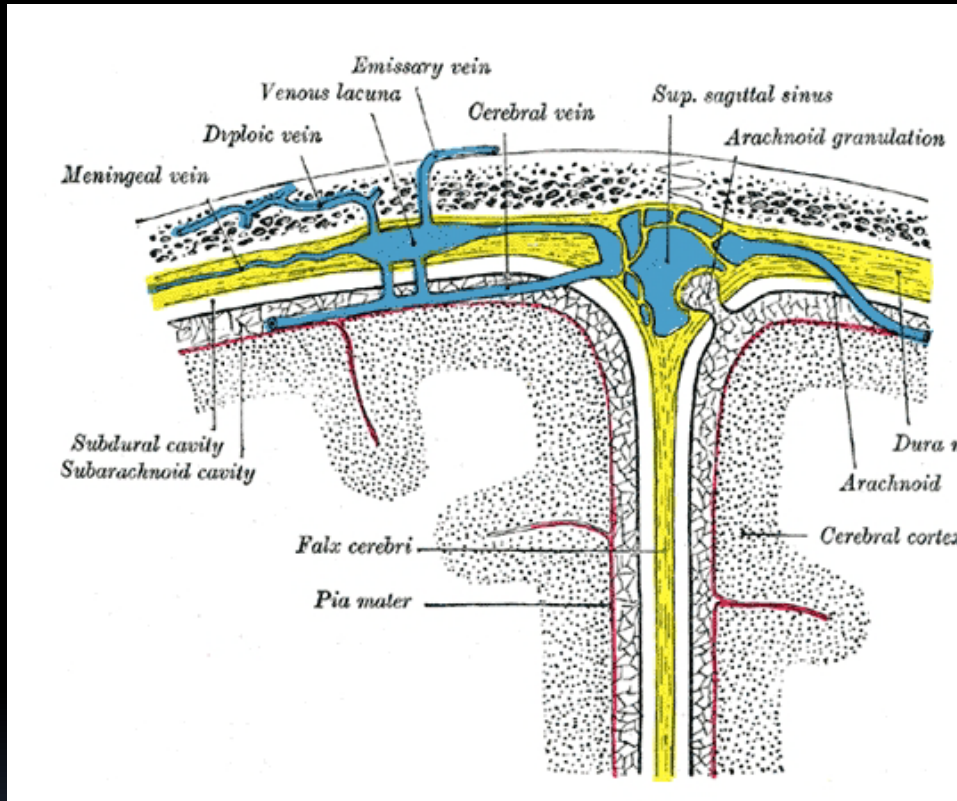
“Shaking-impact syndrome”



BIOMECHANICAL FORCES- Meninges:

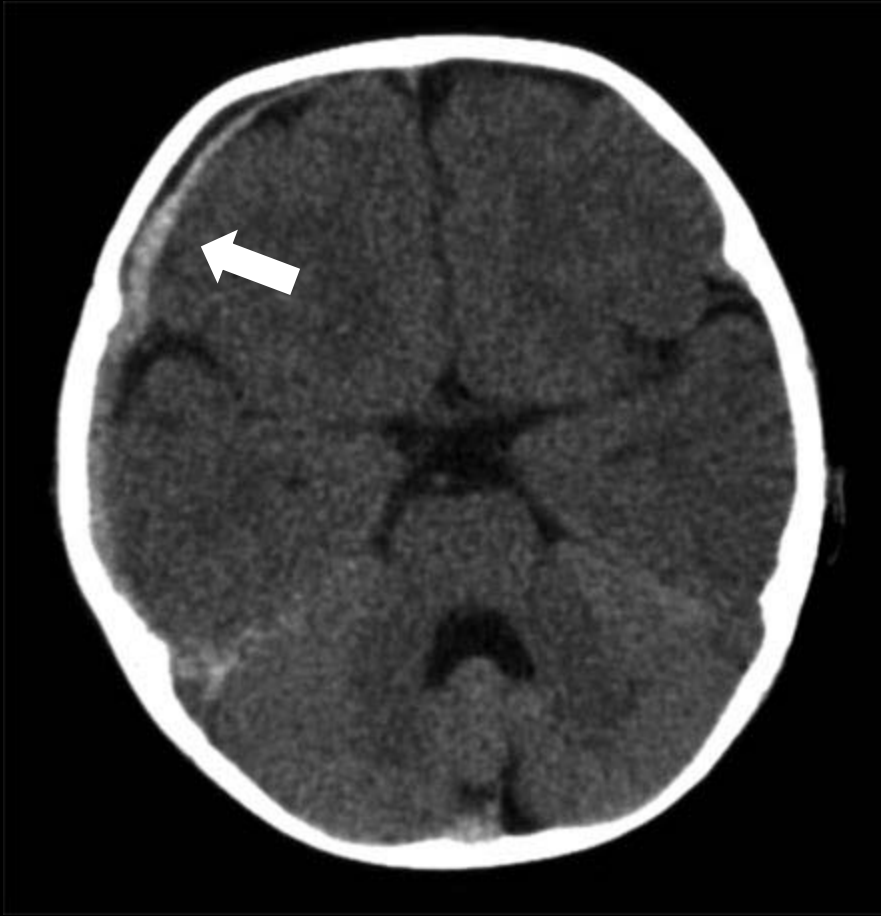


BIOMECHANICAL FORCES- Meningeal veins:



Tearing of meningeal veins →

BIOMECHANICAL FORCES- Subdural hematoma (SDH):

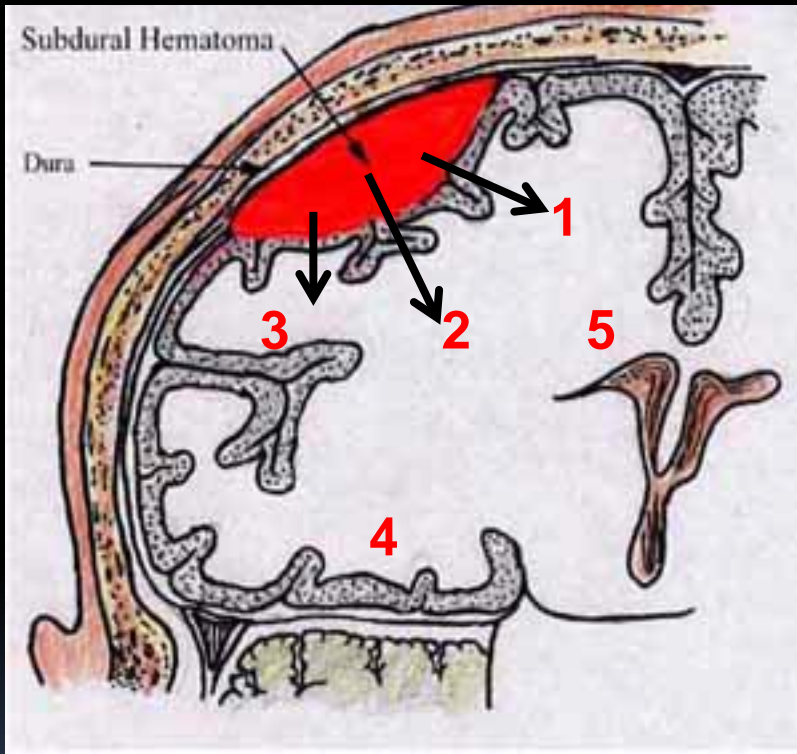


Small SDH



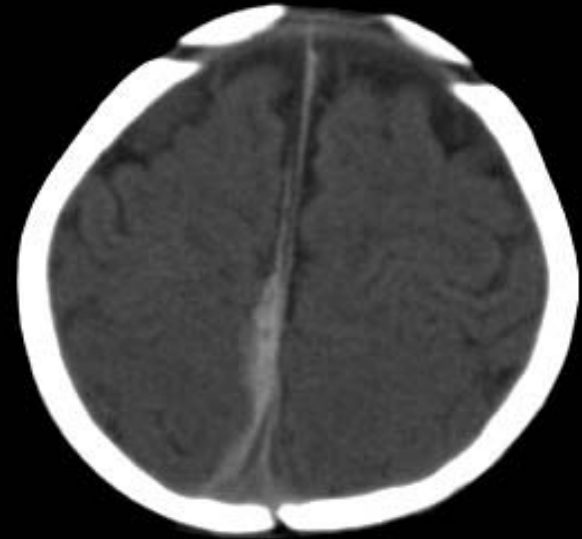
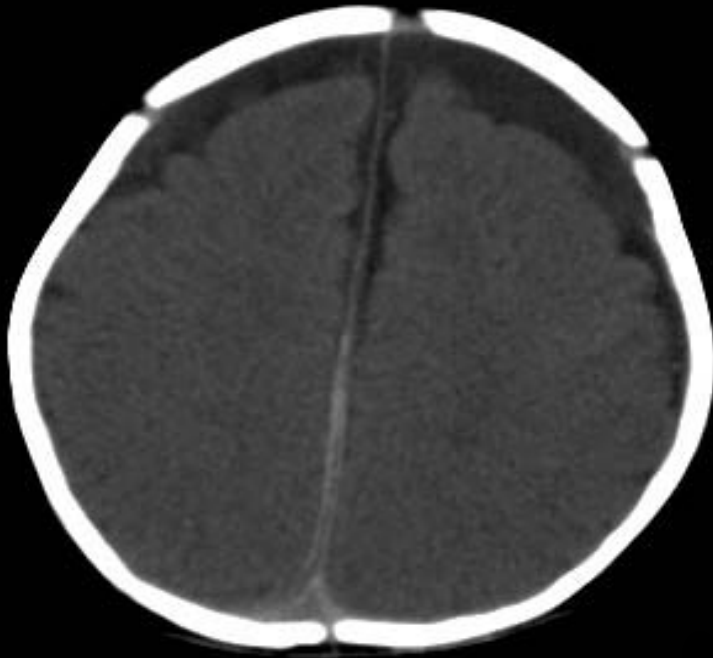
Medium SDH- mass effect

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

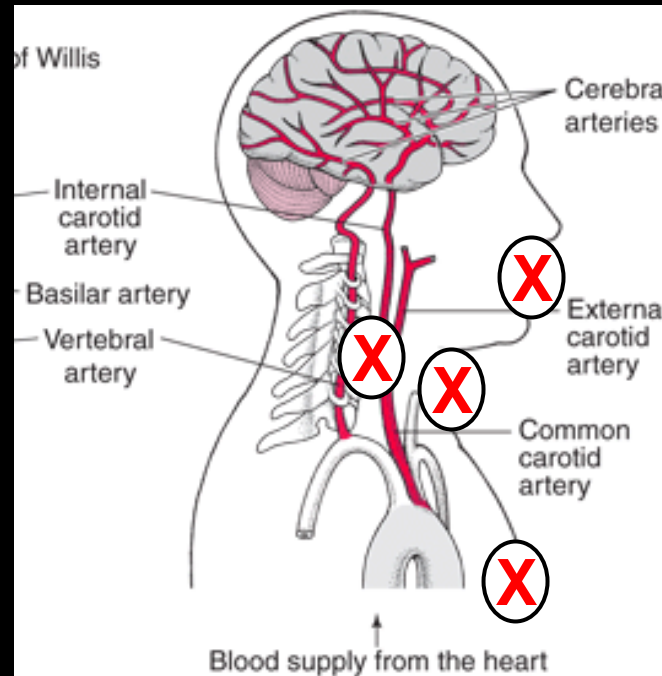
Apnea (restrict chest movement)

Local changes (mass effect of subdural)

“Shaken-impact-suffocation syndrome”- never adopted

Shaking alone can not produce constellation of injuries

METABOLIC FORCES- Hypoxic injury:

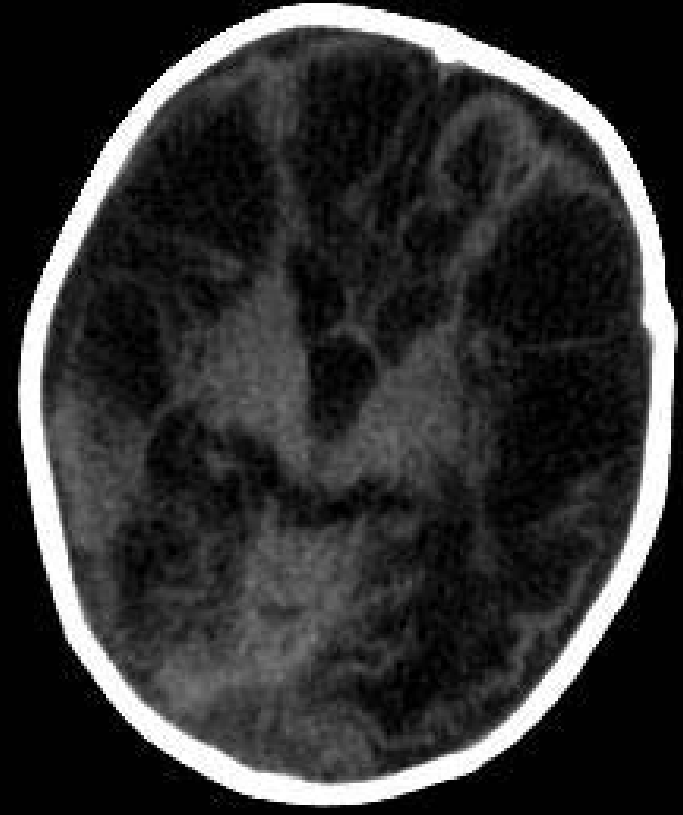


1. Suffocation
2. Choking
3. Strangulation
4. Chest compression

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

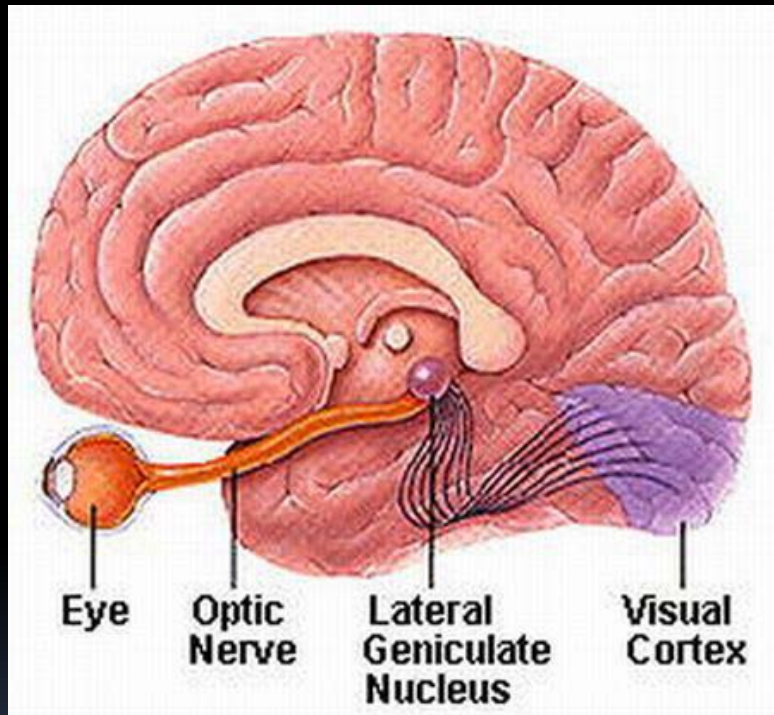
Etiology- Still debated

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

Fragility of developing retinal blood vessels

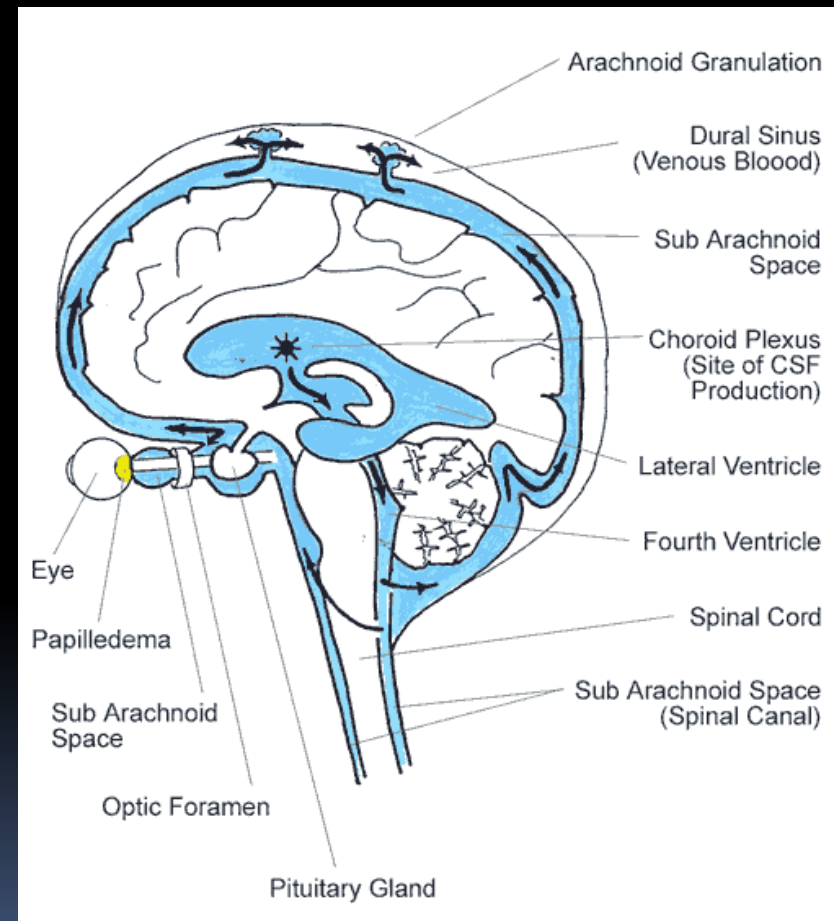
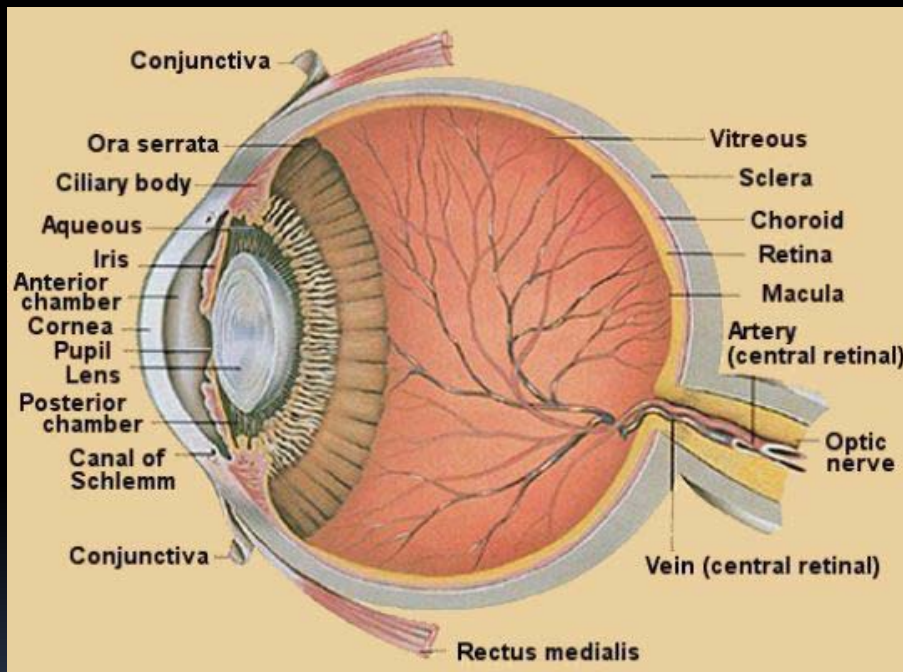
Role of angular deceleration- uncertain

RETINAL HEMORRHAGES- Mechanism:



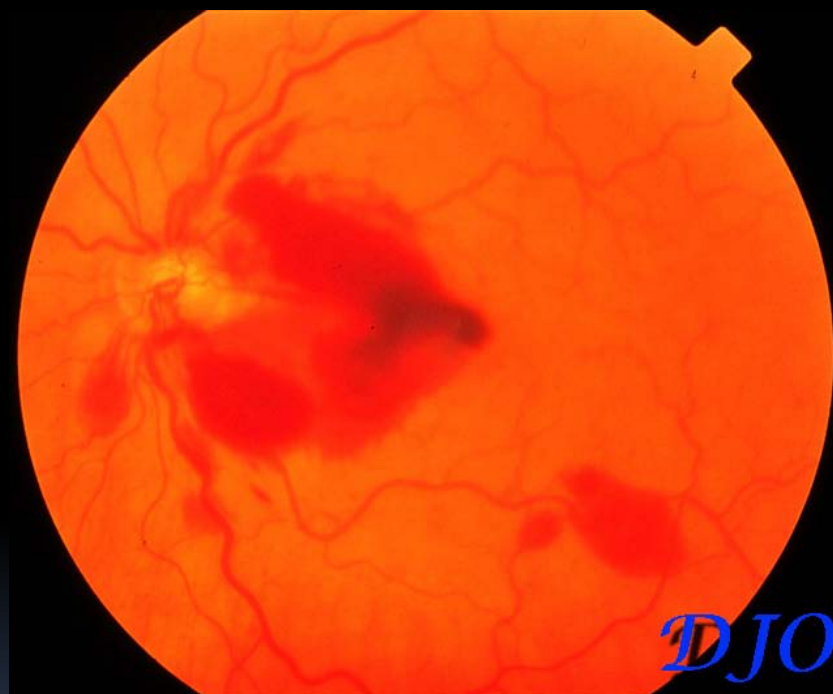
Optic nerve/Eye - part of the brain

RETINAL HEMORRHAGES- 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

SHAKEN BABY SYNDROME:

Subdural hematoma + Retinal hemorrhages + Bone fracture = Child abuse



Subdural hematoma + Retinal hemorrhages = 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 ~~=~~ Child abuse

Medical dogma held for >30 years

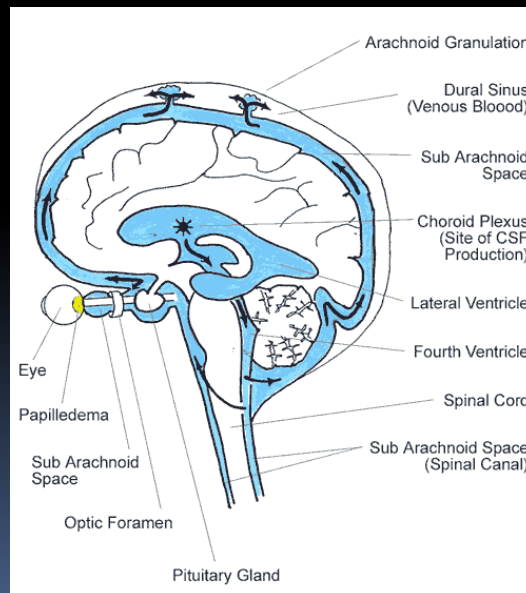
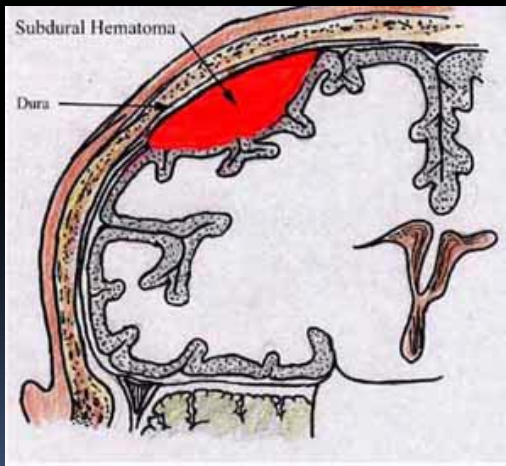
Forces ~~of~~ work and fragility ~~of~~ developing brain =
so unique that two findings together were
incontrovertible

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

Cardiopulmonary resuscitation

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

	<u>SDH</u>	<u>RH</u>
Vaginal delivery	25%	40%
Caesarian section	1-3%	10%

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?

Plunkett J. (2001) Fatal pediatric head injuries caused by short-distance falls. *Am J Forensic Med Pathol* 22:1-12

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?

Bechtel K et al. (2004) Characteristics that distinguish accidental from abusive injury in hospitalized young children with head trauma. *Pediatrics* 114:165-168

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

Vinchon et al. (2010) Confessed abuse versus witnessed accidents in infants: comparison of clinical, radiologic and ophthalmological data in corroborated cases. *Childs Nerv Syst* 26:637-645

n= 45 confessed abuse, 35 public accidents

NEW PERSPECTIVE- Important article:

Plunkett (2001) - reviewed U.S. Consumer Product Safety Commission database for head injury associated with playground equipment

- 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:

Blechtel et al. (2004)- Prospective study of children <2 years admitted for head injury Aug. 2000 to Oct. 2002

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

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

	<u>Abuse</u>	<u>Accident</u>
RH	60%	10%
Unilateral RH	20%	9%
Bilateral RH	40%	1%
Pre-RH	30%	0
Single RH	0	4%
Extends to periphery	27%	0

NEW PERSPECTIVE- Important article:

Blechtel et a. (2004)-

	<u>Abuse</u>	<u>Accident</u>
Seizures	53%	6%
Altered consciousness	53%	10%
Scalp hematoma	7%	51%

NEW PERSPECTIVE- Important article:

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 1st 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:

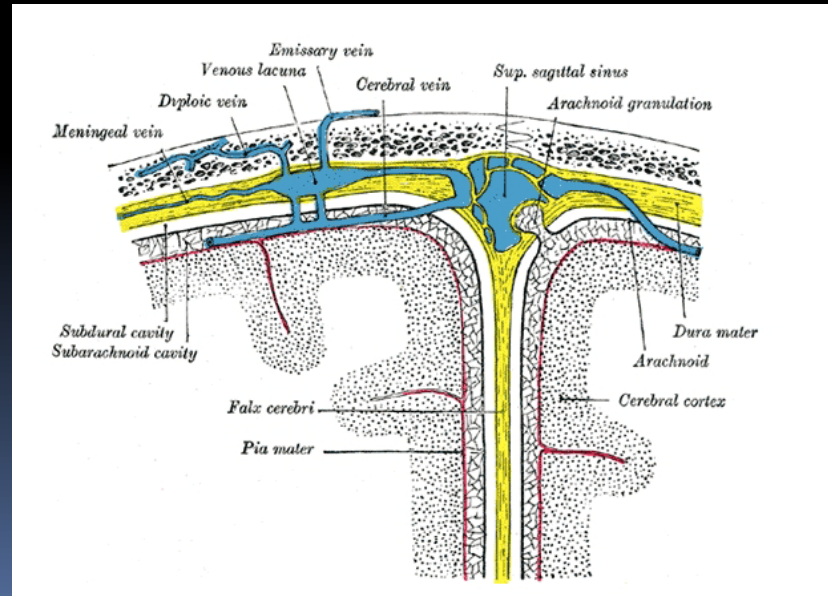
Clinical course:

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:

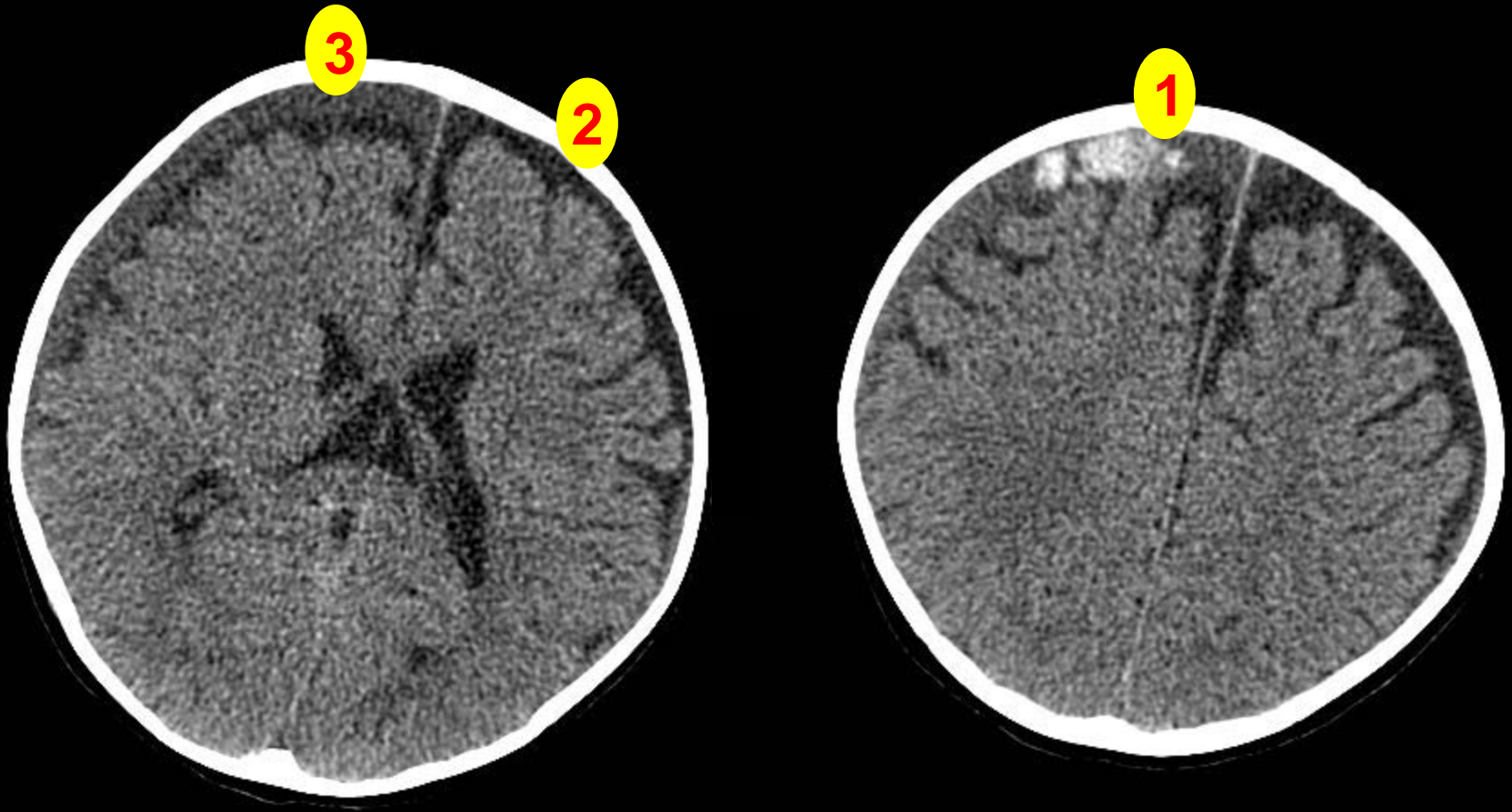


Symmetric extra-axial fluid collection



Right chronic subdural

BENIGN EXTERNAL HYDROCEPHALUS:



“Subdural hematomas of different ages”

BENIGN EXTERNAL HYDROCEPHALUS:

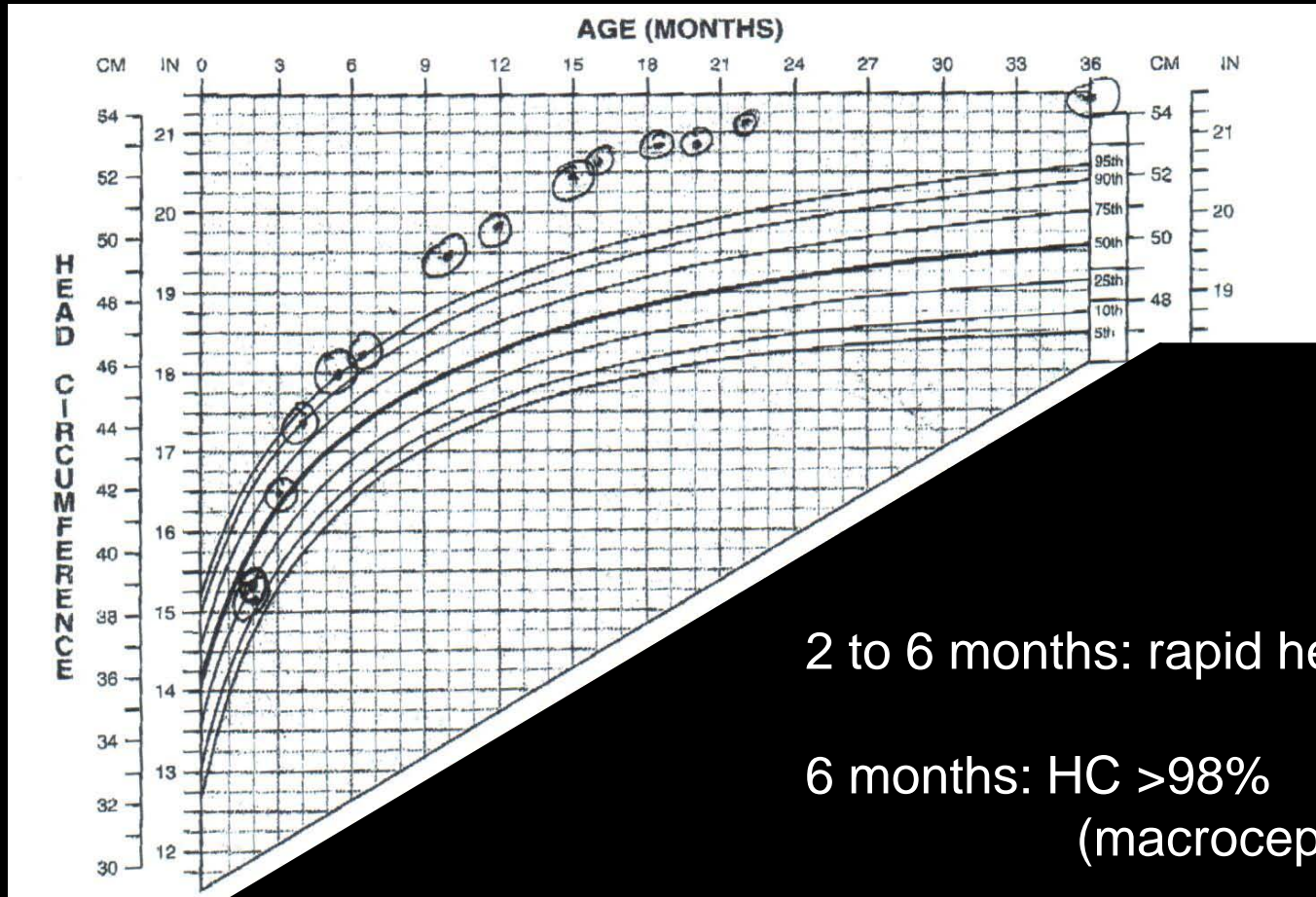
Also, retinal hemorrhages found, R > L

Child removed from the home BUT

Three important facts were overlooked:

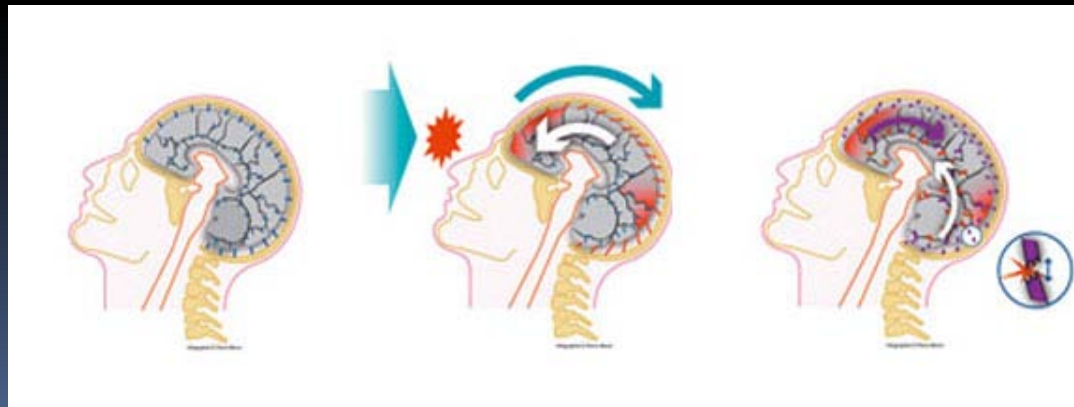
1. Rapid head circumference at 2-6 months of age
2. Dad's head circumference >98%
3. History of forceful rocking behavior, when playful or angry

BENIGN EXTERNAL HYDROCEPHALUS:



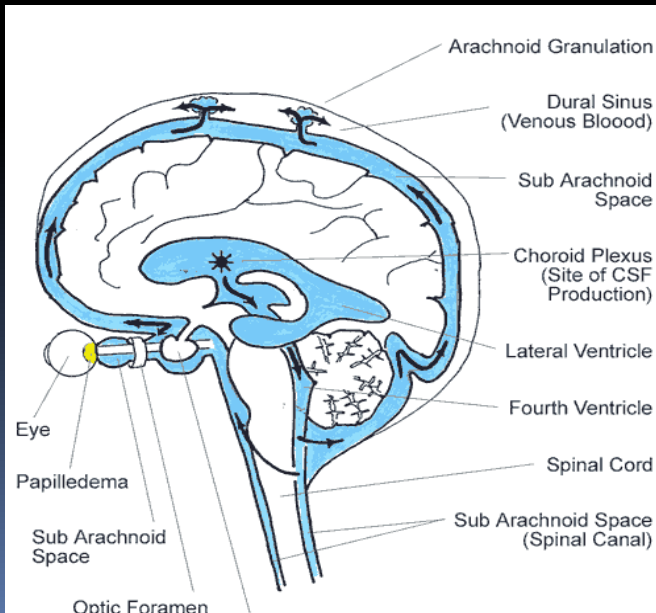
BENIGN EXTERNAL HYDROCEPHALUS:

- 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:

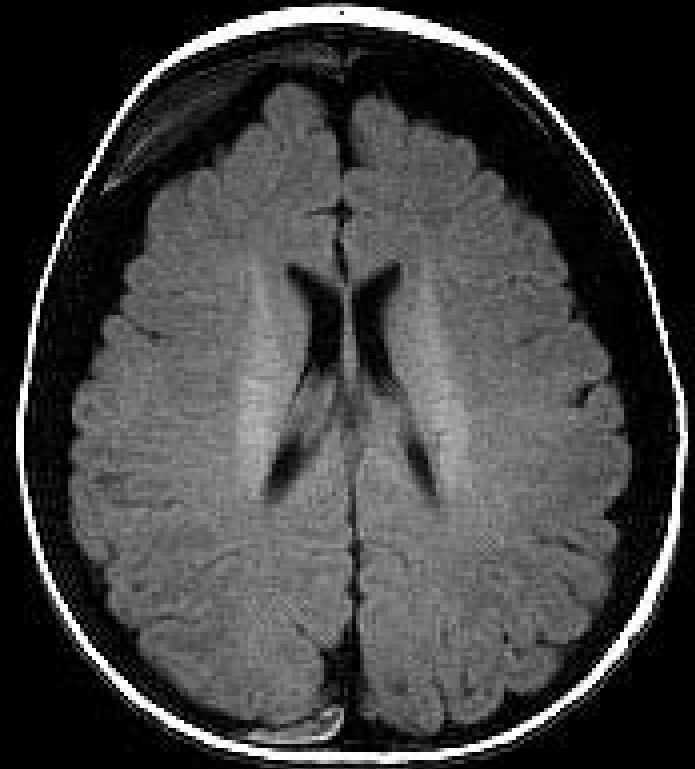
- Consider:**
5. CT scan- Enlarged, symmetric extra-axial fluid collection
 - Known as “Benign external hydrocephalus”
 6. More room for veins to be shaken and torn
 7. Retinal hemorrhages- History of recurrent bleeds
 - this raises intracranial pressure
 - reaches “tipping point” at playground
 - one final “blow” that ruptures vessels



**SUBDURAL HEMATOMA- Distinguishing
subdural from subarachnoid space**



Head CT scan

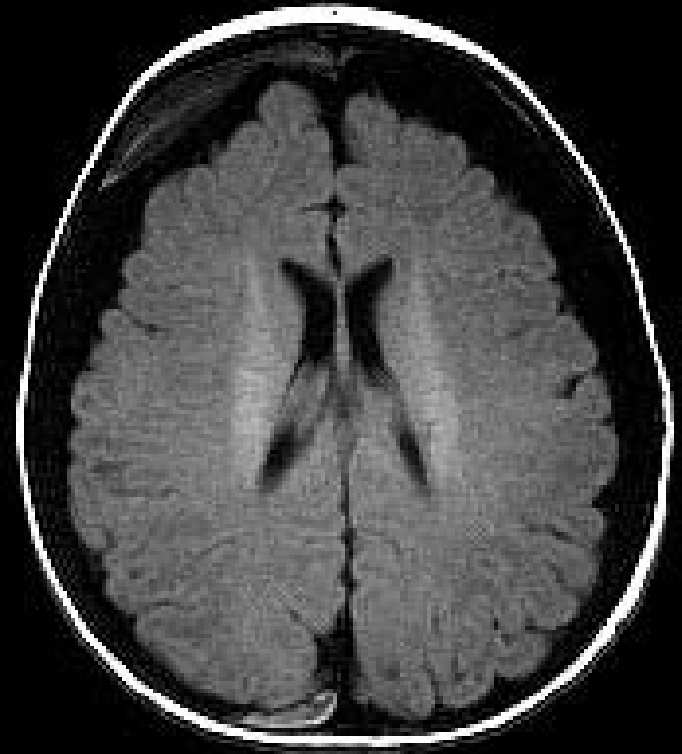


Head MRI scan

**SUBDURAL HEMATOMA- Distinguishing
subdural from subarachnoid space**

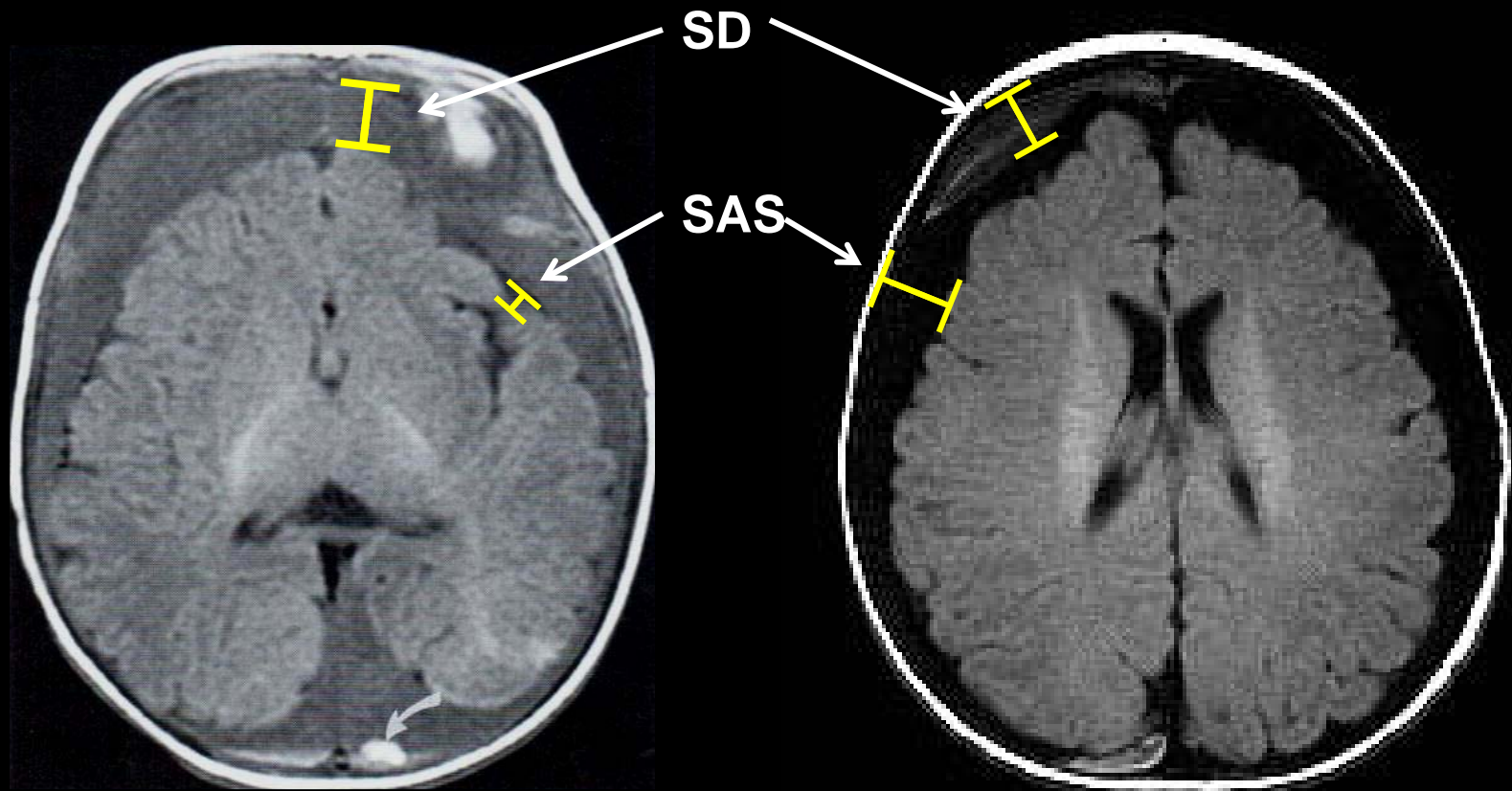


Sagittal view



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:

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

NEW PERSPECTIVE- Important article:

	Abused	Accidental	p-value
<u>Clinical features</u>			
Seizures	31 (68.9%)	5 (12.8%)	<0.001
Somnolence	18 (60.0%)	12 (30.8%)	0.38
Coma	20 (44.4%)	9 (23.1%)	0.04
Deficit	20 (44.4%)	8 (20.5%)	0.02
Raised intracranial pressure	30 (66.7%)	9 (23.1%)	<0.001
Swelling	1 (2.2%)	30 (76.9%)	<0.001
Impact on head	17 (37.8%)	34 (87.2%)	<0.001
Other impact	11 (24.4%)	6 (15.4%)	0.30
Peripheral fracture	12 (26.7%)	2 (5.1%)	0.008
Transfusion	10 (22.2%)	10 (25.6%)	0.71
Life threat	3 (6.7%)	8 (20.5%)	0.060

NEW PERSPECTIVE- Important article:

Table 2 Radiological and ophthalmological findings in the IHI and AT groups

	Inflicted	Accident	<i>p</i>
CT findings			
Subdural collection	37 (82.2%)	17 (43.6%)	<0.001
Mixed density image	39 (90.7%)	17 (53.1%)	<0.001
Brain ischemia	12 (26.7%)	1 (3.2%)	0.0023
Extradural	1 (2.4%)	5 (13.9%)	NS
Contusion	7 (15.6%)	10 (25.6%)	0.25
Fracture	5 (11.4%)	26 (66.7%)	<0.001
Total impact	10 (22.2%)	26 (66.7%)	<0.001
Retinal hemorrhage			
None	7 (15.9%)	29 (82.9%)	<0.001
Mild	3 (6.8%)	5 (14.3%) ^a	NS
Moderate	9 (20.5%)	0	NS
Severe	25 (56.8%)	1 (impact)	<0.001

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

