

Pseudo-Shaken-Baby-Syndrome: A Re-assessment of Shaken-Baby-Syndrome Features

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Abstract

Introduction: Electron microscopy has revealed that sliding of the brain cortex relative to the skull cannot take place without producing contusions, which the SBS definition says are unusual in SBS. There is no natural "Subdural Space" across which sliding can take place and relative movement across the subarachnoid space is restricted by collagen reinforced trabeculae.

The SBS definition can be interpreted as a different syndrome (Pseudo-SBS or P-SBS) involving venous hypertension, having a natural physiological etiology and not involving imposed trauma.

Pre event history: Patients may have a history of poor feeding, vomiting, and lethargy for days or weeks. Extended vomiting is known to cause Mallory-Weiss tears in the mucosa at the junction of the esophagus and stomach. The infant is crying excessively because of pain. Infants may become anemic due to occult bleeding from such sites.

The Triad: Subdural bleeding: SBS is known to be associated with "Forceful" (projectile) vomiting, an indicator of Pyloric Stenosis, in which very high intra-abdominal pressures are involved. Pressurized venous blood may be driven up the vena cavae, into the head including the Superior Sagittal Sinus and bridging veins. It is at this junction that subdural bleeds occur.

Retinal hemorrhages: It occurs in many situations involving extreme intra-abdominal pressure (Valsalva Retinopathy), including forceful vomiting and retching.

Altered consciousness: If the Internal Jugular Vein valves allow jugular venous reflux, "Transient Global Amnesia" may result. Very high pressure may produce permanent brain injury.

Other Features: Diffuse cerebral edema, Macrocephaly etc. result from elevated cerebral venous pressure effusions. The 2:1 M to F infant gender ratio in SBS convictions is accounted for by testosterone accelerated pyloric muscle bulk growth.

Summary: The AAP definition of the physiological features of Shaken Baby Syndrome does define a genuine syndrome, but the cause is physiological not imposed trauma.

Keywords: Shaken baby syndrome; Infants

Introduction

In 1974, Caffey described Whiplash Shaken Infant Syndrome [1] as *The most characteristic pattern of physical findings in the whiplashed infant is the absence of external signs of trauma to the head and the soft tissues of the face and neck and of the facial bones and calvaria, in the presence of massive traumatic intracranial and intraocular bleedings. This is an extraordinary diagnostic contradiction. It is in a large part, responsible for the frequent failures to diagnose subdural hematoma and retinal haemorrhage and the failure to attribute them to manual shaking and whiplashing of the head.*

The terms Non-Accidental Trauma (NAT), Non-Accidental Head Trauma (NAHT), Abusive Head Trauma (AHT) have since been introduced. They are somewhat broader than Shaken Baby Syndrome in that they include other forms of abuse, bruising, broken long bones, skull fractures etc. More recently Paul and Adamo [2] described the current status of SBS.

Traditionally, NAT injury in infants and toddlers has been described as shaken baby syndrome, although the terms inflicted Traumatic Brain Injury and non-accidental head injury have been used in the literature. Only shaken baby syndrome, which has been defined as the triad of subdural hematoma, retinal hemorrhage and encephalopathy suggests

a mechanism of injury in which there is tearing of the bridging veins secondary to shaking. This mechanism remains to be experimentally proven.

The Shaken Baby concept was introduced to explain the observation of severe intracranial injuries in the total absence of extra cranial injuries, the other definitions assume various forms of injury relating to inertia effects but fundamentally rely on the SBS concept for their validation.

SBS is a definite syndrome, it is a medical entity. This article reports a re-assessment of the features reported by the American Academy of Pediatrics Committee on Child Abuse and Neglect in 1993 [3] and in 2001 [4]. The aim was to find if there was an alternative explanation

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in the hope that if so warning signs might also be found to aid its treatment or possibly prevent its development. If such a mechanism were found it would be identified as Pseudo-Shaken-Baby-Syndrome (P-SBS) to signify that it produces the features of SBS but it does not involve mechanical shaking. Throughout this article direct quotes are printed in italics and if from the AAP SBS definition articles, in red. The locations of such quotes in the AAP documents are listed in an appendix. It is suggested that when reading SBS literature in this context, the more neutral term “injury” is substituted for “shaken”, i.e., “Injured child” for “SBS victim”.

Features of the SBS definition

Pre-event history: *In 1972, pediatric radiologist John Caffey popularised the term ‘whiplash shaken baby syndrome’ to describe a constellation of clinical findings which included retinal hemorrhages, subdural or subarachnoid haemorrhages, and little or no evidence of external cranial trauma.* One year earlier Guthkelch had postulated that whiplash forces caused subdural hematomas by tearing cortical bridging veins [5] As early as 1953, Guthkelch, studying ‘Subdural Effusions in infancy’ [6] had reported “*It should be emphasized that infantile subdural effusion is not a rare condition of all surgical conditions of the central nervous system occurring in the first two years of life only spina bifida and hydrocephalus are seen more often than subdural haematoma.*”

Injury threshold: With the coming of electron microscopy it became known that the arachnoid and pia structures are stitched together by collagen fibres crossing the subarachnoid space within trabeculae [7-9]. The brain surface cannot move relative to the skull without tearing these trabeculae off the brain surface. The brain surface injuries so produced are known as contusions. Obviously, if the skull is subject to sufficiently violent movement severe brain damage can be induced. Ommaya et al. investigated the threshold for whiplash injuries [10] by subjecting monkeys to step acceleration while their heads were free to move. They adjusted the impact until 50% of them suffered concussion. Concussion was judged by loss of behaviour responses and loss of corneal and palpebral reflexes. They found there was an all or nothing threshold, below which concussion did not occur. They reported *nineteen animals were considered to have been concussed. Fifteen of these 19 monkeys were found to have visual evidence for macroscopic hemorrhage damage as marked by surface hemorrhages which were primarily subdural in nature. In the nonconcussed group there were 22 in whom adequate post-mortem studies have been completed. Not a single animal in the nonconcussed group showed any macroscopic evidence of brain damage; in particular there were no surface haemorrhages*”. However, the SBS definition [3,4] says *Visible cerebral contusions are unusual, which means that most SBS events do not exceed the stress threshold for mechanical tissue damage.*

Incessant crying: Excessive crying is an important feature of SBS. It is viewed as being the trigger for abusive behaviour by the carer. *“Shaking may seem to be a proportionate response to the tension and frustration frequently generated by a baby’s incessant crying or irritability yet crying is not a legal justification for such violence”.* In SBS cases it had been observed that *“The infant may have a history of poor feeding, vomiting, lethargy or irritability occurring intermittently for days or weeks prior to the time of initial health care contact”.* In P-SBS it is considered that the combination of vomiting and incessant crying suggests Mallory-Weiss tears where the esophagus joins the stomach.

Mallory weiss tears: a source of pain: Tearing of the cardiac orifice of the stomach and the adjoining esophagus during vomiting was discovered by Mallory and Weiss during a study of alcoholic adults in whom vomiting was frequent [11]. Weiss describes their findings as *the*

chief clinical manifestations of these lesions were persistent vomiting and retching it was concluded that pressure changes in the stomach during the disturbed mechanism of the coordinated motor changes which accompany vomiting, and continuous regurgitation of gastric juices over the mucosa of the cardia are the most significant responsible factors.

Gastroesophageal junction mechanics: The upper part of the esophagus descends between the pericardium and the spine in the mediastrum to which it is attached. The esophagus passes through its own aperture in the diaphragm. It is loosely located by elastic fibres in the phrenoesophageal ligament [12]. The phrenoesophageal ligament is a relatively compliant and elastic structure which allows the esophagus to slide relative to the diaphragm. In the adult the gastro esophageal junction moves approximately 2 cm in the cranial direction during a swallow.

Below the diaphragm, the esophagus is attached to the cardia aperture of the stomach. This junction is not a simple end-to-end join, it is more of a splice Z in (Figure 1). The junction follows a zig-zag path around the esophageal and gastric cardia tubes. This junction is known as the Z-line [13]. In the adult this transition region extends over 3-4 cms. Locally, histology of the Z-line marks an abrupt structural change. The esophageal side has a stratified epithelium, but the gastric side has a columnar epithelium. This means that tissue on each side of the Z-line will have different stretch characteristics. A stress concentration will be set up whenever the esophagus is stretched. If the diaphragm pulls the stomach down the gastro-esophageal junction will get pulled down with it, stretching the esophagus longitudinally.

The site of injury: The lower end of the esophagus is surrounded by muscles forming a sphincter [12]. This is not a true sphincter i.e., a ring of smooth muscle cells like the pyloric sphincter it is a collection of muscles with a sling-like action which combine to form a sphincter-like action. Normally this sphincter only has to withstand a small pressure drop for which this arrangement is adequate.

Figure 1a represents the normal situation and Figure 1b represents the situation early in a vomiting event. In Figure 1b the diaphragm has descended. When the diaphragm is maximally contracted (maximum tension) the lungs get sucked down. Some authors describe this as a “deep breath” [14] but this is not a normal inspiration its purpose is compression of the abdomen under the control of the vomiting centre. The abdominal walls are tensioned to form a container in which to generate maximum expulsive pressure on the abdominal contents. Normally the lower esophageal sphincter is closed unless eating or drinking occurs. However, with the diaphragm and abdominal muscles at maximum tension the intra-abdominal pressure will be very high, forcing the lower sphincter open. Chyme at high pressure can then force its way into the esophagus and dilate it. This will stretch the walls of the stomach cardiac orifice and the attached local esophagus. The walls of the stomach are lined with rugae which can unfold to accommodate stretch in the associated muscle layers. The mucosa in the walls of the esophagus is lined with plicae [13]. Plicae cannot unfold, and so are less elastic. The join is locally abrupt and known as the Z-line. Such joins of materials of different elasticities are known to produce stress concentrations when stretched. When the Z-line region is excessively stretched by the lumen pressure the mucosa may tear, Mallory-Weiss tears [15-17]. Once the mucosa is torn it will heal in a few days if undisturbed, but if another surge occurs the wound will be re-opened. In the meantime any reflux will bring acid stomach contents into contact with the wound leading to erosion, as Weiss found [11].

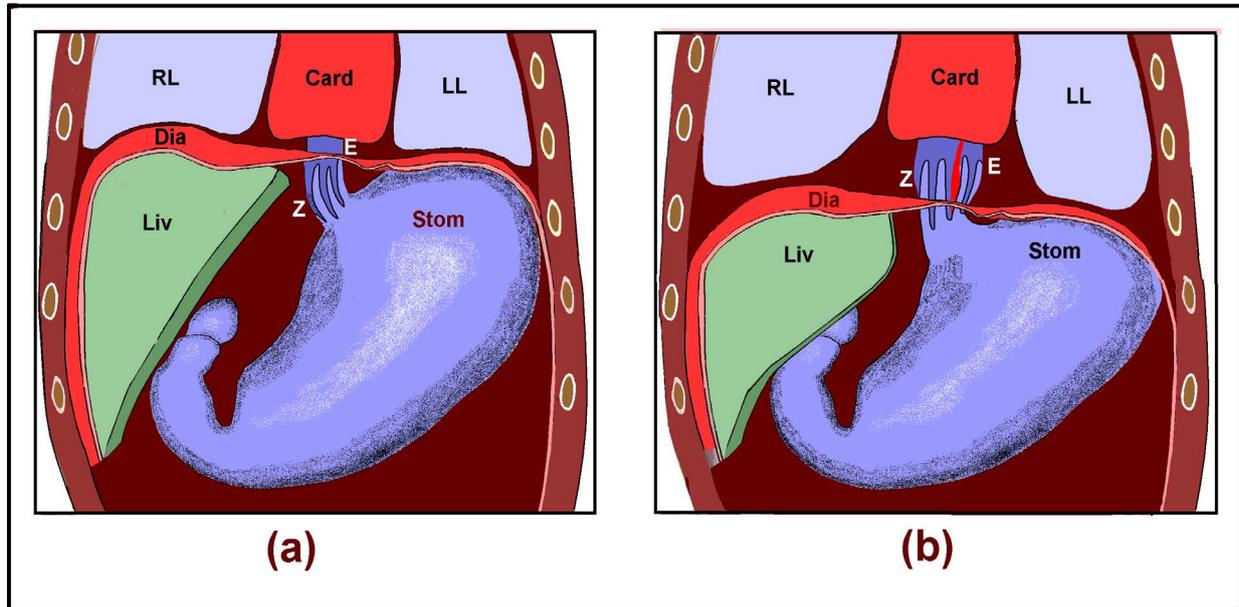


Figure 1: Organ configuration; (a) in normal quiet breathing, and (b) during vomiting. Card: Cardiac Sac; Dia: Diaphragm; E: Esophagus; Liv: Liver (Part of); LL: Left Lung; RL: Right lung; Z: Z-line. The esophagus and the stomach cardia are joined by interdigitated extensions. This results in the gastro-esophageal junction following a Zig-Zag path around their extensions, the Z-line.

Medline Plus Medical Encyclopedia [15] says “A Mallory-Weiss tear occurs in the mucus membrane of the lower part of the esophagus or upper part of the stomach, nearby where they join. The tear may bleed. Mallory-Weiss tears are usually caused by forceful or long term vomiting.” Slow bleeding from the surface of a tear would immediately be mixed with food in the stomach fundus and so not be noticed. This would account for the observation in SBS cases that “*The shaken infant is often mildly to moderately anemic.*” In SBS the incessant crying is considered to be the cause (trigger) of injuries. In P-SBS it is the result of the disorder. The infant is crying or screaming because it is in pain.

Intra-cranial venous hypertension: On the last page of his last publication on SBS [1], Caffey describes the vomiting as forceful. Forceful vomiting is associated with some form of obstruction of the alimentary canal, typically a sign of pyloric stenosis. The pyloric muscle may go into spasm, or grow so thick that its lumen cannot become patent even when the muscle is completely relaxed. This causes the brain to call for maximum effort from the diaphragm and abdominal muscles to clear the blockage. The excessive intra-abdominal pressure produced drives venous blood up into the head [18]. The P-SBS concept offered here is that the intracranial injuries presenting in the syndrome can arise from Cerebral Venous Hypertension (CVH) such as is produced during the violent vomiting and retching associated with Pyloric Stenosis [19]. It would be impracticable, and probably unethical, to attempt to measure the intra-abdominal pressure in an infant during forceful vomiting, but non-bilious projectile vomiting in an infant with pyloric stenosis is known to throw vomit several feet across the room [20], requiring really high intra-abdominal pressures.

The pylorus: physiology and function: One form of alimentary obstruction, particularly relevant in the first few months of post-natal life is Pyloric Stenosis. The Pylorus organ is situated between the stomach and the intestine (duodenum). It has two components, a sphincter and

a canal shown red in (Figure 2).

The canal muscular wall (Figure 3) is much thicker than that in the cardiac parts of the stomach. Its mucous membrane is in longitudinal folds along its whole length.

During the first stage of digestion (in the stomach) the pyloric sphincter is held closed while the canal contracts vigorously and repeatedly to reduce food items to a paste. This process is often referred to as “grinding”, but mammals have no grinding mechanism. Birds and reptiles have a specialised grinding organ, the “gizzard”, separate from the stomach [21] but the mammalian pyloric canal, (Figure 3), is streamlined and lubricated. The mammalian system is optimised to produce high viscous shearing forces on any lumps in the path of powerful jets of digestive fluid driven from the pyloric canal back into the stomach [22].

Pyloric development: This stripping action requires powerful systolic action by the pyloric canal musculature, but at birth the pylorus muscle is still immature, longer but thinner compared to its adult form (Figure 4) [23,24]. Alarotu, in his survey of the anatomy and physiology of the pylorus [25] comments that “*The circular muscle system of the stomach has been observed to develop in the human embryo at the 23-41 mm stage, but the longitudinal muscle layer does not develop until the 41-75 mm stage. However, segments in the longitudinal muscle are still absent at birth, and they do not achieve full development until about one year subsequently.*”

Lewis [23], commenting on the form of the stomach in 10 mm human embryos observes that “*Since the incisure in the adult is perhaps twice as far from the cardia as from the pylorus, it is evident that the pars pyloric are relatively long in early stages.*” Postnatally, the pyloric muscle has to grow rapidly to prepare for post-weaning “stripping” of food lumps, but if it gets too thick too soon there may be little room for chyme to pass on into the intestine even when the muscle is fully

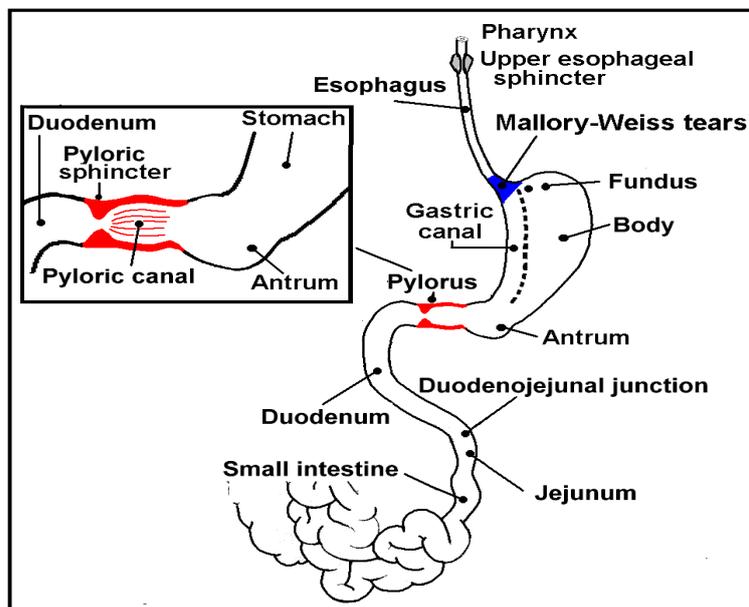


Figure 2: The alimentary canal. The position of the Pyloric Sphincter and Pyloric Canal are shown in red. The site of Mallory-Weiss tears is shown in blue.

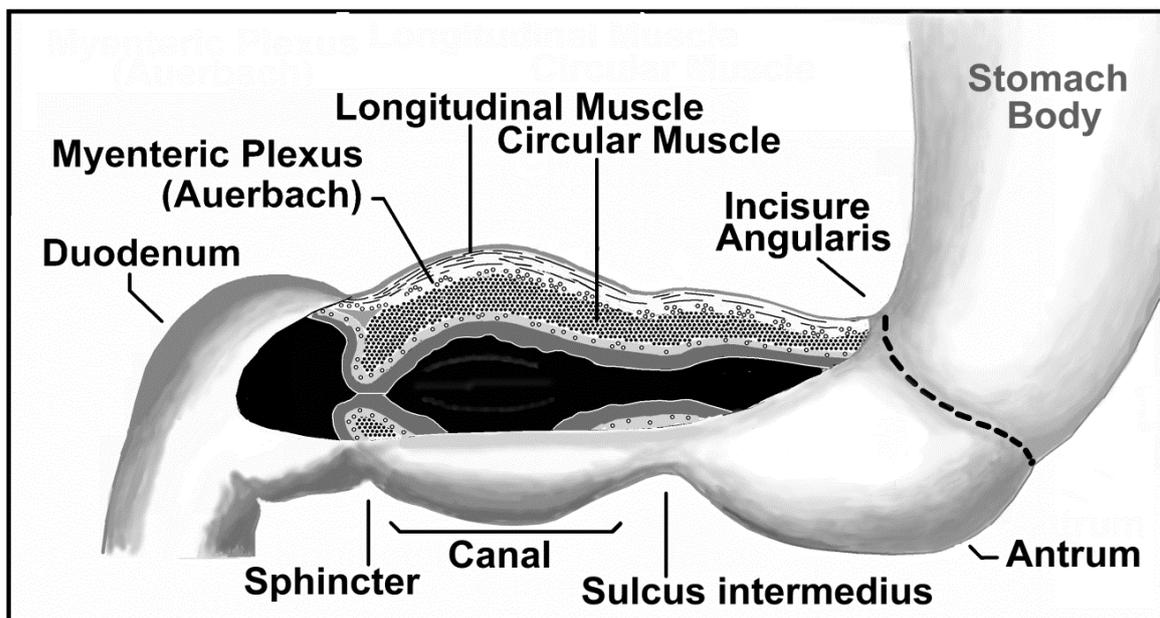


Figure 3: The pylorus. The pylorus has two components, the Sphincter and the Canal. They contract independently but cooperatively.

relaxed. The pylorus is said to have become stenosed. Challa studied myenteric plexuses and the smooth muscle of the pylorus, obtained by biopsy at pylorotomy in 5 infants with hypertrophic pyloric stenosis, using electron microscopy [26]. They found that “*The maturation and development in infantile hypertrophic pyloric stenosis are not impaired for this disorder. The presence of small immature neurons and large mature neurons suggests that neuronal maturation and development are not impaired as previously reported. Except for hypertrophy of the circular smooth muscle layers no specific alterations were found in muscle fibres of the pylorus.*”

Cerebral venous hypertension during violent vomiting

During vomiting, the diaphragm, (D) in Figure 5 and abdominal muscles (AM) compress the abdominal contents (spleen, liver, and all the veins) against the Pelvic Bowl. The resultant high intra-abdominal pressure will expel blood up the inferior vena (IVC), and retrograde up the superior vena cava (SVC). Some protection of the extremities is given by valves in the femoral, brachial, and jugular veins. However, in man, the jugular valves have evolved to optimise low forward resistance at the

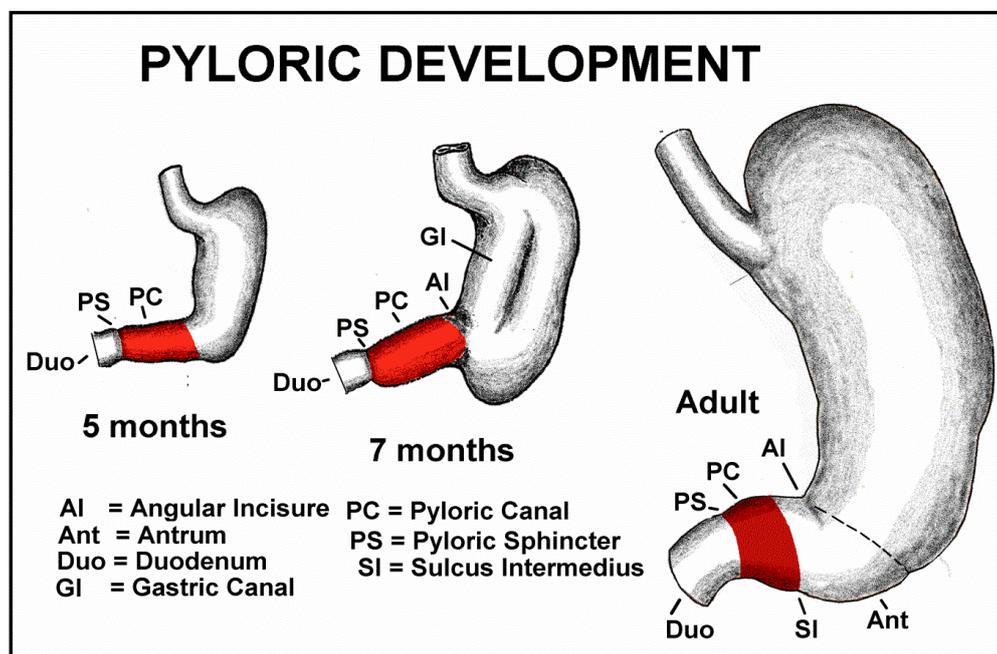


Figure 4: The pylorus develops as a specialised section of the alimentary canal in utero. It does not attain adult proportions until several months post natal. Postnatally it becomes proportionally shorter and thicker.

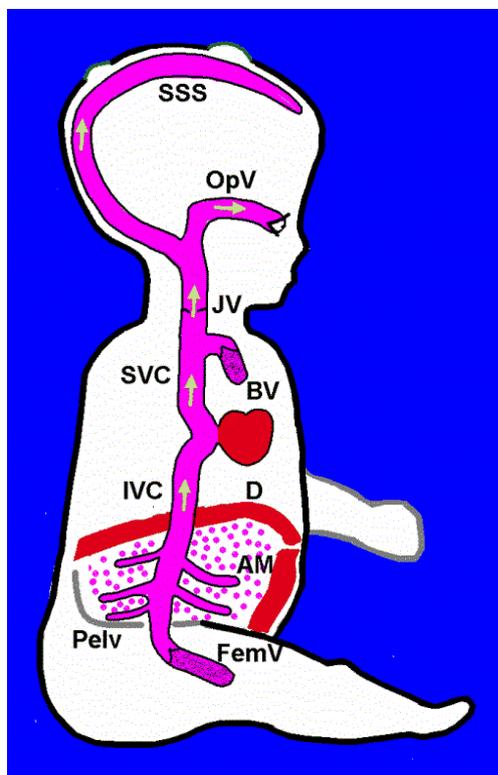


Figure 5: Structures involved in Cerebral Venous Hypertension (CVH) generation. AM: Abdominal Muscles; BV: Branchial Vein Valves; D: Diaphragm; FemV: Femoral Vein Valves; IVC: Inferior Vena Cava; JV: Jugular Veins; OpV: Ophthalmic Vein; Pelv: Pelvic Bowl; SSS: Superior Sagittal Sinus.

expense of reverse pressure protection capacity [27]. There are no valves in veins within the cranium. If the jugular vein valves are inadequate, pressurised blood will enter the brain's venous system (Figure 6). If this happens various forms of injury are to be expected, to the eyes, bleeding into the Subdura, and general cerebral malaise.

There are two circulations in each eyeball, the Retinal and the Uveal [28]. The retinal layer is supplied by the retinal artery and vein which run along the centre of the Optic nerve these are the vessels that can be seen on the inner surface of the retina in ophthalmology. The rest of the eyeball is supplied by the Uveal circulation. These are branches of the ophthalmic circulation and enter the eyeball without passing within the optic nerve. Both the retinal vein and the Ophthalmic vein drain into the Cavernous Sinus. (Sinuses are grooves in the skull in which veins run. The infant skull is too thin to have grooves, so strictly speaking, in the infant the term refers to vessels which subsequently will have sinuses develop around them. The Cavernous Sinus is a complex structure of anastomosing vessels. Each Cavernous Sinus is drained via its inferior and superior Petrosal sinuses into their Sigmoid or transverse Sinuses. Normally, blood from each sigmoid sinus drains out via its Jugular Vein into its Brachiocephalic Vein. There are usually valves in the Jugular veins near the Jugular or Brachiocephalic junction to prevent reflux in the event of Brachiocephalic vein pressure surge. Within the brain there are no further valves. In the event that these valves are weak or absent any reflux driven by a venous pressure surge will travel throughout the vessels shown in (Figure 6). Reverse flow travelling along a transverse sinus to the Torcular, and up the Superior Sagittal Sinus will pressurise the Bridging Veins it normally drains, causing Subdural Hemorrhage. That travelling back into the Retinal and Ophthalmic veins will cause Retinal Hemorrhages, and widespread venous pressure rises will cause altered mental function. Thus R-SBS indicates that the elements of the triad are not independent variables, they are manifestations of a common variable, venous hypertension.

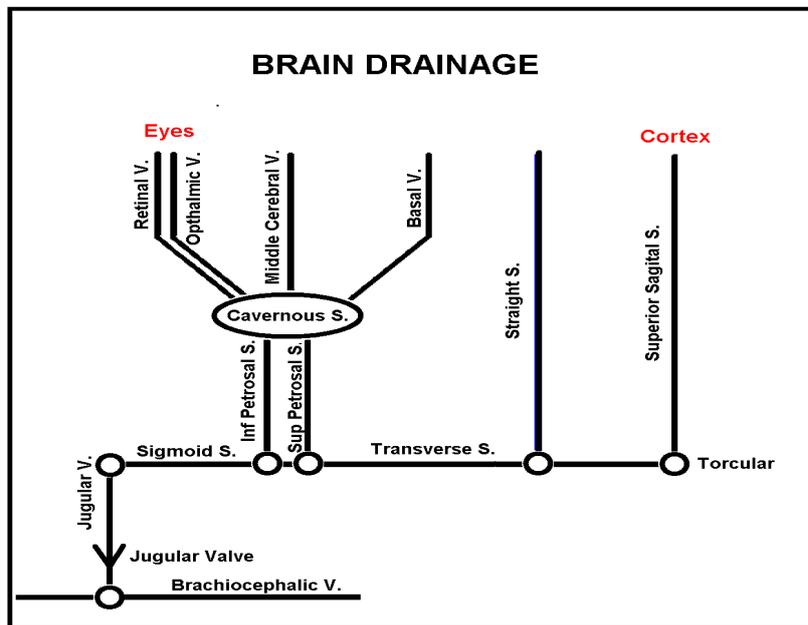


Figure 6: The brain's venous system. There are no venous valves within the cranium. If a Jugular Vein Valve fails under pressure all the major brain veins will be subject to SVC pressure surges.

Clinical Relevance of Physiological Features

The concept of the Shaken Baby Syndrome (SBS) [3] was created before electron microscopes had revealed that the arachnoid and pia maters were stitched together by trabeculae reinforced by collagen fibres (Figure 7).

Hence a brain cortex cannot slide relative to the arachnoid mater as required by the SBS hypothesis, without causing noticeable damage (contusions), but trabeculae are not mentioned in the SBS definition.

Subdural Bleeds (primary element of the triad)

The SBS foundation document [4] alleges that *subdural hemorrhage caused by the disruption of small bridging veins that connect the dura to the pia arachnoid is a common result of shaking*. This depends on shaking being able to move the brain without causing contusions. The SBS document says *visible cerebral contusions are unusual*.

Where could sliding take place; Subdural? or Subarachnoid?

The "Subdural" Space

In the 1970s when the SBS concept was created it was thought that the inward order of membranes in the meninges was Dura, Fluid-Space, Arachnoid, Pia whereas it is now known that the order is Dura, Arachnoid, Fluid-Space, Pia. The 1970s concept of a subdural space (Figure 8) still forms the basis of the SBS concept today.

In 1991 Haines surveyed the literature on the subdural space. He found 42 publications concerning, or descriptions of, the subdural space from contemporary (1991) textbooks or atlases in gross anatomy, histology, and neuroscience [31,32]. He quotes Romanes (1986) that *"The arachnoid is separated from the dura by a bursa-like, capillary space (the subdural space) containing a film of fluid. This forms a sliding plane where movement is possible*.

Haines, summarising his findings at that time says:

A representative survey of the extant teaching literature reveals that the majority (36/42) state categorically that a subdural space is present between the arachnoid and dura. In a minority of this sample (6/42) the probable existence of a subdural space is clearly qualified or specifically denied. (A true potential space is one that may be created without disrupting the normal structure or functional integrity of the tissues involved in creation of the space [33]. So at that time there was general agreement that a subdural space, or at least a potential space, existed. But with the coming of Electron Microscopy it was found that there was no pre-existing subdural space, it was a preparation artefact.

It was found that whereas most of the dura was toughened by copious collagen fibres (Figure 9), the lower boundary (then known as the Dural Border Layer, DBL) was devoid of collagen. This layer is so different from the main dura that it is in effect a separate structure in its own right. It will be referred to as the Subdura [34]. Thus there is a sandwich of Dura, Subdura, and Arachnoid. The Dura and the Arachnoid are reinforced with collagen but the Subdura between them is relatively weak. When sufficient stress is applied, the Subdura is torn apart (Figure 9), remnants of the Subdura being found on the Dura and Arachnoid.

Summarising, Haines says *"The appearance of the so-called subdural space is the result of tissue damage and is not due to the expansion of a patent (temporarily obliterated) pre-existing space. Consequently, this space is neither "actual" nor "potential"; it is in fact, non-existent in the normal situation. The so-called subdural hematomas are actually dural border hematomas"*.

However the term "Subdural" is widely established and may continue to be used with the new meaning of within, or close to, that part of the Dura recognised as the "Subdura". P-SBS considers the possibility that "Subdural" bleeds result from the transient venous hypertension occurring during forceful vomiting. The relevant layers of tissue lining the inside of the skull are shown in Figure 10. They are

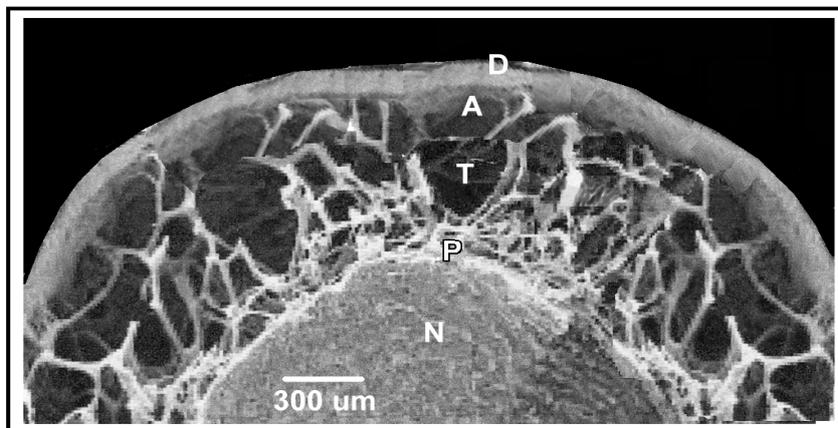


Figure 7: Cross-section Electron micrograph of a human optic nerve. Prepared from figures in reference [8]. The optic nerve is actually an extension of the brain reaching out to the eye. In man the optic “nerve” (N) contains over a million myelinated axons [29]. Like the brain it is covered by an extension of the dura (D) and arachnoid mater (A). Between the arachnoid and the pia maters is a CSF filled space, through which collagen reinforced trabeculae (T) stitch the arachnoid and pia maters together.

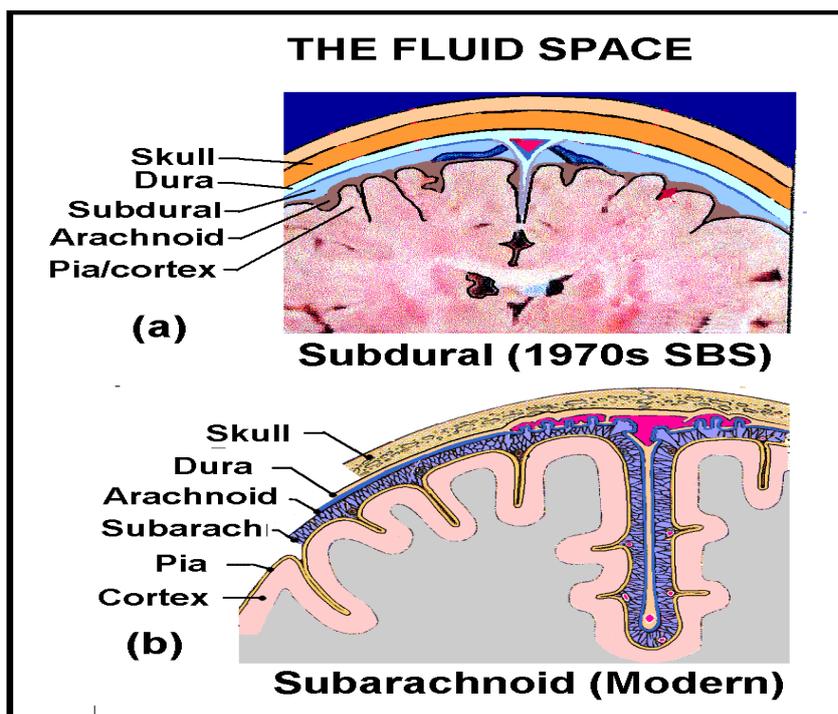


Figure 8: Fluid spaces as represented in the SBS concept (a) and modern text books (b). In SBS the arachnoid mater is assumed to lie close on the brain (figure 8a) Drawn from Figure 2 in reference [30]. In SBS (a) a fluid space is thought to lie between the dura and the arachnoid mater and there is no subarachnoid space. Electron microscope studies, (b), have shown there is no subdural space, but there is a fluid filled space between the arachnoid and pia maters, i.e., subarachnoid.

known collectively as the meninges. The Superior Sagittal Sinus (SSS) is a major collecting sinus (vein) running from front to back just under the skull. The dura mater, which lines the inside of the skull, is mostly very tough, being extensively reinforced by collagen fibres, but its inner surface layer (Subdura) is devoid of reinforcement. Below the dura border is a further membrane, the Arachnoid (Gr spiderlike) with its “cob-web” like tangle of very fine cords and sheets, “trabeculae” on its underside [35]. These are too thin to be seen on ultrasound or MRI.

Collagen fibres within these trabeculae stitch the arachnoid, across the fluid filled subarachnoid space, to the innermost lining layer, the Pia Mater or cortex. So, sliding cannot take place at the Dura or Arachnoid boundary or in the subarachnoid space without causing contusions. Bleeding from the bridging veins must be by some other mechanism than relative brain or skull relative movement. Any alternative explanation must account for both the generation and the location of these bleeds.

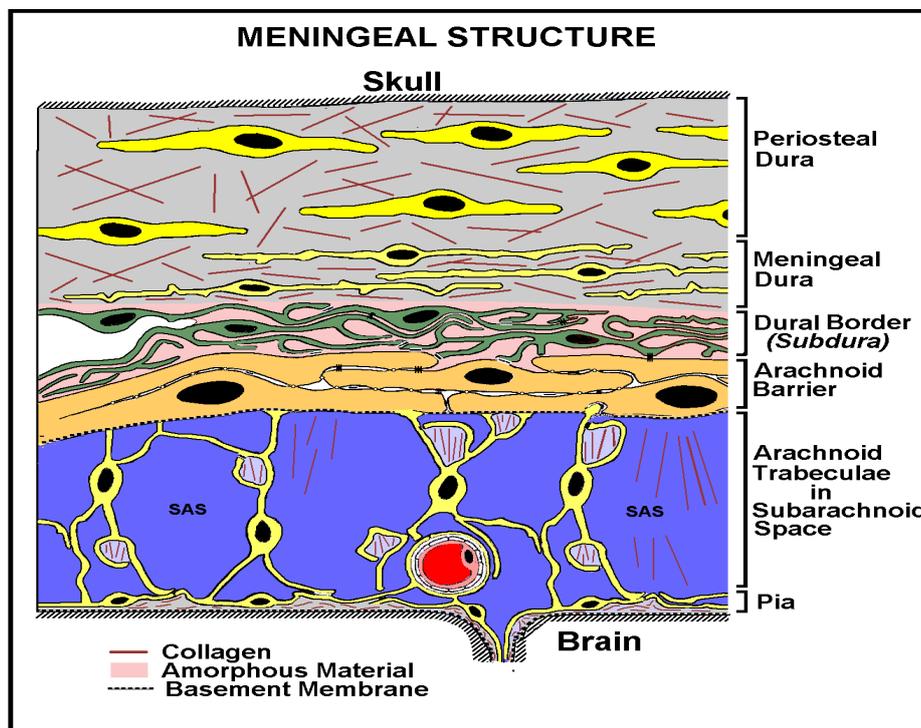


Figure 9: Developed from Figure 1 in reference [33]. A specialised layer of fibroblasts is found at the dura-arachnoid junction, characterised by flattened fibroblasts, no extracellular collagen, extracellular spaces and few cell junctions. It is structurally weak and can be torn apart by the main Dura and Arachnoid membranes, as on left.

Subdural bleeding

Venous blood from the brain cortex normally reaches the SSS through the bridging veins. Bridging veins get their name from the fact that they accept blood from the cortical veins on the brain cortex and carry it across the subarachnoid space (Figure 10a), and up through the meninges into the Superior Sagittal Sinus (SSS). If, during violent vomiting, high pressures reach the SSS, blood will flow backwards into the bridging veins stretching them. Where the bridging veins pass through the soft Subdura they may leak water under this pressure (Figure 10b). This water will be trapped between the impervious arachnoid barrier layer below and the impervious dura mater above. It can only enter the Subdura, causing it to swell, and eventually split (Figure 10b). As the splitting of the subdura proceeds the parts of bridging veins attached above and below the split get pulled apart stretching the part of the vein between them (Figure 10c). Eventually the vein wall will be torn. Then any subsequent venous pressure surges will act directly in the split, bringing blood to form a hematoma (Figure 10d). This mechanism was shown experimentally in pigs by Orlin [36].

This combination of “subdural” haemorrhages without contusions matches the SBS statement that “subdural” hematomas are common in Shaken Baby cases but contusions are “unusual” [4]. However, in experimental shaking Ommaya found the opposite. Contusions in the

cortex could occur in the absence of subdural bleeds [10]. This indicates that the SBS mechanism must differ from the shaking mechanism occurring in Ommaya’s experiments.

Retinal hemorrhages (2nd element of the triad)

The SBS defining document declares: *In 75% to 90% of cases, unilateral or bilateral retinal haemorrhages are present.* In P-SBS Retinal Injury is believed to be due to raised intra-abdominal pressure, known as Valsalva Retinopathy “The term Valsalva retinopathy refers to hemorrhage in and around the macula in response to a sudden rise in intra-thoracic or intra-abdominal pressure. The increased intravenous pressure is felt to be transmitted to the retinal circulation with resulting retinal capillary rupture” [37]. It is recognised in adults in many everyday situations. Compressive thoracic injury [38], Weight lifting [39] Coughing, vomiting [40], Valsalva maneuver [41] and Aerobic exercise [42]. In most cases normal sight returns in a few months. In SBS it is thought that *Retinal and vitreous haemorrhages and nonhemorrhagic changes, including retinal folds and traumatic retinoschisis, are characteristic of shaken baby syndrome.* In P-SBS they are thought to result from episodes of excessive cerebral venous pressure [43]. In SBS *Evidence of prior child abuse is common. Specific evidence of previous injuries (e.g., old intracranial hemorrhages) from shaking episodes is found in about 33% to 40% of all cases.* In P-SBS old intracranial hemorrhages are considered evidence of previous venous hypertensive episodes.

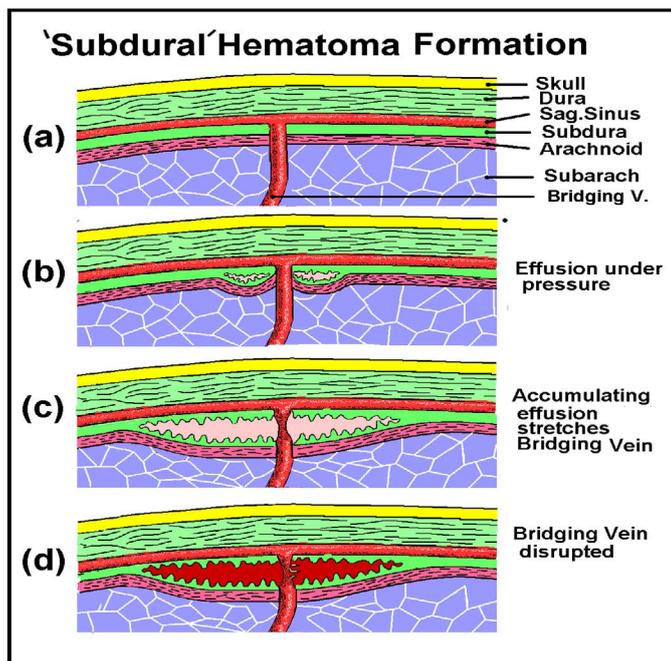


Figure 10: Production of a "Subdural" hematoma by cerebral venous hypertension surges (a) Normal configuration of the meninges, (b) water effusing through the bridging vein walls under pressure begins to accumulate in the Subdura. (c) Distension of the Subdura by water driven through vein walls during cerebral venous pressure surges. (d) Local vein wall failure allows blood at current venous pressure to enter the split Subdura and act directly, increasing distension.

Altered consciousness (3rd element of the triad)

Temporary loss of consciousness in infants resembles "Transient Global Amnesia" (TGA) in adults [44]. Transient Global Amnesia is a benign condition characterised by sudden anterograde amnesia for less than 24 hours without further focal neurological deficit. The postulated mechanism is thought to occur through a Valsalva like mechanism. Supporting evidence for this was contributed by three ultrasound studies which found a significantly higher prevalence of retrograde internal jugular vein flow, Inner Jugular Vein (IJV) incompetence, during a Valsalva manoeuvre in TGA. Nedelmann studying jugular valve closure in adults [45], concluded that cerebral venous hypertension caused by intensive and prolonged Valsalva strain and facilitated by jugular valve insufficiency plays a significant role in the pathogenesis of TGA. TGA is normally considered a disorder of the elderly but it would also account for the temporary "Blank Stare" reported during the period preceding an SBS event [46].

Extra-Triad Observations

Pylorospasm

Spasmodic contractions of the pylorus sometimes occur in infants, usually between 2 and 12 weeks of age. Pylorospasm is characterised by failure of the smooth muscle fibres encircling the pyloric canal to relax normally [24]. In P-SBS the effect during spasm will be the same as in pyloric stenosis. Cohen [47] concluded that pylorospasm may mimic hypertrophic pyloric stenosis.

Macrocephaly (Large heads)

Guthkelch noted in 1953 [6] that occasionally the parents of infants presenting with subdural bleeding had noticed enlargement of the head. More than half showed a significant degree of enlargement of the

head (that is a circumference greater by 1 in. (2.5 cm) Miller and Miller found that Infants involved in SBS convictions also commonly had large heads [48]. In SBS it is thought that the significance of macrocephaly is that it would make infants more vulnerable to shaking, i.e., it is part of the cause of the problem. In P-SBS, macrocephaly is considered to result from a form of external hydrocephalus [19]. When the sutures between the developing cranial bones are stretched the dura beneath them secretes materials that accelerate skull growth [49]. A surge in venous pressure will produce a burst of effusion through vessel walls and a surge in CSF volume. This will stimulate head growth rate, resulting in macrocephaly. Inspection of clinical histories indicates that accelerating head circumference growth may give warning of imminent danger [50, 51].

Gender bias

In a modern textbook [24], Congenital Hypertrophic Pyloric Stenosis is said to affect approximately 1 of every 150 male infants but only 1 of every 750 female infants. The International Pediatric Endosurgery Group (IPEG) quote a 4:1 M/F ratio [52]. The University of Rochester Medical Centre say Pyloric Stenosis is four times as common in males as in females [20]. Guthkelch, studying infantile Subdural Effusions of Infancy [6], noted an M/F incidence ratio of infants presenting of 5:1. He quotes Ingram and Matson, who found a preponderance of males over females in the ratio of 5:3.

Male infant preponderance in convictions for SBS: the influence of testosterone

Miller and Miller [48] conducted a literature search from 1996 to 2006 for infants with Subdural Hemorrhage who also were the subject of successful SBS convictions. They found 62.6% were male, an M/F ratio of roughly 2:1. P-SBS offers the explanation that this results from

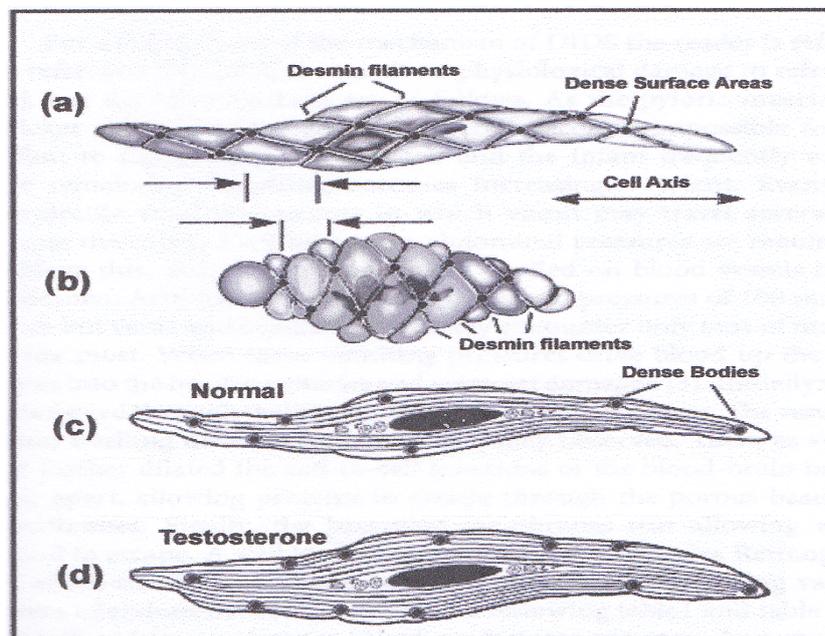


Figure 11: Typical smooth muscle cell. Desmin filaments form a string-bag surrounding each smooth muscle cell. The bag is attached to the cell surface at “Dense surface areas” When the actomyosin elements shorten (b) they pull the dense areas along the cell axis and the cell contents bulge between the desmin filaments. If the cell is pulled while it is relaxed it can be stretched until the desmin, and extracellular collagen (not shown) are tight. Testosterone accelerates actomyosin production (d) making cells fatter but not longer.

the action of testosterone on production of contractile proteins in the developing pylorus muscle.

Visceral smooth muscle cells use actin/myosin filament (contractile protein) “motors” like voluntary muscles, but in a different configuration that allows greater foreshortening. Smooth muscle cells are protected from overstretching by filaments of desmin around and within individual cells, and collagen and reticulin fibres between cells, forming a “safety-net” (Figure 11) [53].

When relaxed the smooth muscle cells can be stretched freely until these safety nets are pulled tight, then they strongly resist further stretch (Figure 11a). Testosterone accelerates production of actomyosin proteins within the cells but has no direct effect on the desmin and collagen safety-net. Cells get fatter, but grow in number and length at the normal rate (Figure 11d). Hence in boys the circumference of the pylorus tends to grow at the normal rate, but contractile protein bulk growth is accelerated by testosterone. The accelerated pyloric muscular bulk development means that boys will be likely to reach stenosis earlier than girls. It has been proposed that this would account for the remarkable (>2:1) preponderance of boys in cases of macrocephaly, subdural hematoma, and in convictions for SBS [22].

Conclusion

Physiologically the SBS definition is consistent with a hydro-mechanical condition. Excessive intracranial edema and hemorrhages are caused by episodes of cerebral venous hypertension. Significant features are:

- Screaming in the period preceding an event is not the cause of events, it is the result of Mallory-Weiss tears during previous violent vomiting.

- A “Subdural Space” does not normally exist; it is an artefact of tearing through the weak layer in the dura where the Dura and Arachnoid meet. Previously known as the “Dural Border Layer” it is suggested that its importance justifies its being known as “The Subdura”. Terms such as “Subdural Hemorrhage” can then be retained with the revised meaning of “through or near the Subdura”
- The brain cannot move as whole without causing contusions which are known to be unusual in SBS. The soft brain can move by deforming, but the absence of contusions indicates that the cortex remains attached to the meninges [54].
- The significant gender bias suggests that testosterone driven pyloric stenosis is a dominant cause of P-SBS. Other mechanisms also exist. For instance in utero mal-development of intracerebral veins may raise intracerebral vein pressures sufficiently to produce cerebral edema and haemorrhages [55].
- The elements of the triad (Subdural Bleeds, Retinal Hemorrhages, and Altered Consciousness) are not independent variables; they result from a common venous hypertension episode.
- P-SBS is a purely physiological occurrence independent of whether a career is present or not.

While it has been assumed that the source of the problem lay outside the body (imposed trauma), little attempt has been made to look for internal mechanisms, or for warning signs and preventative measures. Research into patterns of head growth may be very rewarding.

Appendix

These are a collection of quotes from the AAP 2001 definition of SBS that appear in this article. Their location is indicated by page

number p.N followed by column number c.1 or 2. This refers to their position as down loaded from PEDIATRICS vol 108 No 1 July 2001

- **Presumption of abuse:** *Although physical abuse in the past has been a diagnosis of exclusion, data regarding the nature and frequency of head trauma consistently support the need for a presumption of child abuse* p.206 c1.
- **Frustration:** *Such shaking often results from tension and frustration generated by a baby's crying or irritability, yet crying is not a legal justification for such violence.* p.206 c. 2.

Previous events

- **History:** *The infant may have a history of poor feeding, vomiting, lethargy, and/or irritability occurring intermittently for days or weeks* p.207 c1.
- **Non-specific signs:** *These nonspecific signs are often minimised by physicians or attributed to viral illness, feeding dysfunction or infant colic.* p. 207 c.1.
- **Prior abuse:** *Evidence of prior child abuse is common. Specific evidence of previous cranial injuries (e.g. old intracranial hemorrhages) from shaking episodes is found in about 33% to 40% of all cases* p.207 c.1.

Findings on examination

- **Altered consciousness:** *The brain injured infant may be convulsing, may have altered consciousness, may not be able to suck or swallow, and may be unable to track with eye movements, smile, or vocalise.* p.207 c.1
- **Anemic:** *The shaken infant is often mildly to moderately anemic* p.207 c2.
- **Retinal haemorrhages:** *In 75% to 90% of cases, unilateral or bilateral retinal haemorrhages are present* p.207 c.2
- **Contusions:** *Visible cerebral contusions are unusual* p.208 c.1.
- **No external injury:** *Externally visible injuries are often absent* p.206 c.1.
- **Bridging veins:** *Guthkelch had postulated that whiplash forces caused subdural hematomas by tearing cortical bridging veins.* p. 206 c1.
- **Bridging veins:** *Subdural hemorrhage caused by disruption of the small bridging veins that connect the dura to the pia arachnoid is a common result of shaking.* p.208 c.1.
- **Previous injuries:** *Shaken baby syndrome is unlikely to be an isolated event. Evidence of previous cranial injuries (e.g. old intracranial hemorrhages) from shaking episodes is found in about 33 to 40% of all cases.* p.207 c1.
- **Axonal injury:** *Diffuse axonal injury is common* p.208 c.1.
- **Characteristic retinal injuries:** *In 75% to 90% of cases, unilateral or bilateral retinal haemorrhages are present. Retinal and vitreous haemorrhages and nonhemorrhagic changes, including retinal folds and traumatic retinoschisis, are characteristic of shaken baby syndrome.* p.207 c.2.
- **Meningitis:** *At times, the clinical signs suggest meningitis, and a spinal tap yields bloody cerebrospinal fluid.* p.207 c2.
- **Xanthochromic CSF:** *Xanthochromic fluid should raise the suspicion of cerebral trauma that is at least several hours old.* P.207 c2.

- **MRI v CT:** *Sato et al. have demonstrated a 50% greater rate of detection of subdural hemorrhage using MRI, compared with CT* p.207 c.2.

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