

## The Legacy of John Caffey: Shaken Baby or Pyloric Stenosis?

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### Abstract

The American Academy of Pediatrics' 1993 definition of Shaken Baby Syndrome (SBS) refers to John Caffey's 1972 publication describing a syndrome including retinal hemorrhages, subdural and/or subarachnoid hemorrhages. In his 1974 follow-up paper emphasis is on possible predictive factors in the infant's history. Particularly significant are bulging fontanelles and "Forceful" vomiting. Bulging fontanelles suggest excessive intra-cranial pressure, and forceful vomiting suggests pyloric stenosis. It is this invitation to consider the possible involvement of vascular hypertensive factors that is his legacy, the subject of this paper.

The SBS hypothesis assumes that the brain "floats" in the skull tethered only by the bridging veins. Actually, the apparently vacant subarachnoid space is filled by a mesh of collagen reinforced fibres, called "trabeculae". These are too fine to register on Ultrasound or MRI screens. Importantly, it is these trabeculae (not the bridging veins) that position the brain in the skull. They also locate and support the bridging veins. These trabeculae are not mentioned in the SBS literature, rendering the SBS hypothesis invalid in its present form.

Forceful vomiting produces extreme venous hypertension which can induce Valsalva Retinopathy (bleeding in the eyes), and tearing (splitting) of the subdural compartment, leading to subdural hemorrhage. Pyloric stenosis occurs in 1 in 150 male infants, but only 1 in 750 females, a M/F ratio 5:1. Male preponderance is also found in convictions for "Shaken Baby Syndrome", 62.6%. (M/F 2:1). Here, this is attributed to testosterone accelerated production of contractile proteins in pyloric smooth muscle cells, causing them to swell and block the lumen earlier than in females.

Caffey kept records of possible associations of factors, whether he could see the significance or not. His SBS legacy is the recognition that pressure, due to a naturally occurring disorder, rather than imposed trauma, could cause the injuries presently attributed to shaking.

**Keywords:** Shaken baby; Macrocephaly; Bridging veins; Venous hypertension; Pyloric stenosis; Gender bias, Subdural hematoma; Arachnoid

### Introduction

John Caffey was born in 1895, the same year that Rontgen discovered x-rays. He volunteered for a post-WWI tour of duty in Europe, and stayed on to work for the American Relief Administration. On returning to Michigan he completed his residency in medicine and became a pediatrician on the staff of The Babies Hospital in New York.

He was put in charge of the hospital radiology department, and pediatric radiology became his life's work. Dr Caffey was a founding member of The Society for Pediatric Radiology in 1958.

Thorne Griscom [1] described Caffey as a true scholar in the best sense; he loved to dig to the bottom of whatever was being considered, and he was devoted to the truth. He often said "it's not who is right, it's what is right". He did not put much value on opinions, he wanted to know the facts. This was reflected in a special file he started keeping when at the Babies Hospital. He called it his "learning file". It contained any observations that caught his attention whether they were understandable or not. Some of these latter seem to have been incorporated in his last (1974) paper on Shaken Baby Syndrome and stimulated the research reported in this article.

In 1993 the American Academy of Pediatrics, Committee on Child Abuse and Neglect, issued a statement defining Shaken Baby Syndrome; Inflicted Cerebral Trauma [2]. They referred to John Caffey's studies.

*"In 1972, pediatric radiologist John Caffey popularised the term "whiplash shaken baby syndrome" to describe a constellation of clinical findings in infants, which included retinal haemorrhages, subdural and/*

*or subarachnoid hemorrhages, and little or no evidence of external cranial trauma".*

Actually, in 1972, the then current term was "Battered Babe Syndrome", but it was typical of Caffey that in 1972 he would not use it [3]. Although he clearly was convinced that the intracranial injuries were the result of shaking by the carer, "Shaken" is not in fact mentioned in the title of this paper. Caffey entitled his paper "*The Parent-Infant Traumatic Stress (PITS) syndrome*". He thought that such titles as "Battered Babe Syndrome" could spark a premature bias against accused parents before adequate medical and legal investigations could be made. He explains "*The term battered child may be crucially unjust when used before the guilt of the parents has been established legally. PITS syndrome is a fair name; it accuses no one. It does indicate the causal, emotional, social and economic stresses which plague the mother or her substitute*".

This was characteristic of the man. All his life he remained convinced of what had been done (shaking) but he did not want his article to bias a jury, before a trial, in deciding whether it had been done. It was not until after Guthkelch had postulated that whiplash

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forces caused subdural hematomas by tearing cortical bridging veins [4] that Caffey relented and entitled his 1974 [5] follow up paper.

*The Whiplash Shaken Infant Syndrome: Manual Shaking...* These two papers, 1972 and 1974 form the basis of this present article. Direct quotations are shown in italics.

### Caffey’s 1972 Summary of the Current Status of “Battered Babe Syndrome”

Caffey commented that although Battered Babe Syndrome had been known for 25 years its true incidence was unknown. He summarized current knowledge (in 1972) as:-

#### Cause

The cause of the trauma was willful assault principally by mothers (90%), but in less frequency by fathers, siblings, other relatives, parent-substitutes, and others of several kinds (baby sitters, “boy-friends” of the mother, cleaning women, infant nurses, delivery boys, etc.)

#### Perpetrators

Perpetrators of the assault were characteristically of normal intelligence and represented all races, creeds, in all cultural, economic, social and educational levels and were distributed proportionately in all parts of the country.

#### Victims

Victims were usually normal infants, but the incidence of abuse was higher in provocative, deformed, premature, multiple-birth, adopted and step children. They were characteristically not neglected or deprived of material needs; they are almost always clean, well fed and well clothed. Many infants, victims of subdural hematoma, might have been shaken by adults, either for discipline or in play, without damaging long bones.

#### Caffey’s ‘74 paper

Caffey’s ‘74 paper is strongly influenced by Guthkelch’s postulate that whiplash forces could tear cortical bridging veins where they joined the Sagittal Sinus surfaces [4]. Guthkelch had published photographs of lacerations at these sites. As by 1974 it was commonly accepted that shaking was the only possible explanation of the subdural hematomas found in this condition, Caffey was now free to define the condition as “Shaken Baby Syndrome”,(SBS). This was universally accepted as more appropriate than “Battered Babe Syndrome”. He incorporated SBS in the title of his 1974 paper:-

*The Whiplash Shaken Infant Syndrome: Manual Shaking by the Extremities with Whiplash-Induced Intracranial and Intraocular Bleedings, Linked With Residual Permanent Brain Damage and Mental Retardation.*

In this paper emphasis is on the infant, the career is barely mentioned.

Caffey noted [5] that Ingram and Matson had found subdural hematomas from all causes to be lesions essentially of the first year of life, with a peak-age incidence of 6 months. Their histories disclosed a high incidence of such infantile complaints as vomiting, 48%; hyperirritability, 41%; infections, 39%; stupor, 32%; history of birth trauma, 26%; and of postnatal trauma, 20%. There was an absence of a history of trauma of any kind in 54% which was considered to suggest that whiplash shaking might be the cause in many patients.

### Comparison

Comparing the two articles, the ’72 paper recommends various measures to reduce stresses on mothers, the ’74 paper gives a list of factors in the infant itself that might warn of impending fatal injury.

In the 1972 paper all injuries are assumed to be caused by mechanical force, there is no mention of pressures or any hydrodynamic features. The ’74 paper introduces bulging fontanelles, forceful vomiting, congested and edemous optic nerves, etc. Caffey points out the plasticity of the infant skull. He interprets it as a factor increasing the movement of the brain during shaking. He quotes Guthkelch’s observations of bridging cerebral veins being torn from their attachments. He quotes Ingraham and Matson’s findings of fevers in 57% of subdural hematoma cases. He notes the incidence of enlarged heads. He quotes Ingraham and Matson that subdural hematomas were essentially lesions of the first year of life. He realized that “*These facts indicate that many features of post-traumatic subdural hematomas are not satisfactorily explained or understood; namely, the nature of the primary causal trauma in more than half of the cases, the exact causal mechanisms of the combination of subdural and intraocular bleedings, or the vulnerability of the affected structures and tissues to whiplash stresses.*” He directed that “careful diagnostic consideration “should be given in infants showing this syndrome”.

### Subdural bleeding

In SBS subdural bleeding was attributed to tearing of cortical bridging veins at the Dural/Sagittal Sinus junction. It was believed to result from excessive tension in veins arising during shaking of the infant by the career. The skull was assumed to rotate around the brain, stretching these veins until they pulled out of their attachments [4,5]. Bridging veins are attached to the inside of the skull (the dura mater) at one end, and to the surface of the brain (Pia Mater) at the other. The brain had been assumed to float freely in Cerebrospinal fluid (CSF), restrained in position by the bridging veins. If the infant was shaken, these veins would attempt to pull the brain to follow the skull rotation. If the shaking was violent enough it was believed that the tension in these bridging veins might become sufficient to pull them away from the Dura allowing haemorrhaging and leaving torn tissue at the site. But there are doubts as to whether these veins are not too elastic to convey the necessary tension.

Squier describes a method of examining the bridging veins [6]. *The skull is opened by parasagittal cuts lateral to the superior sagittal sinus. By carefully lifting the midline bony strip the bridging veins can be visualized,* (Figure 1). In human infants the distance across the

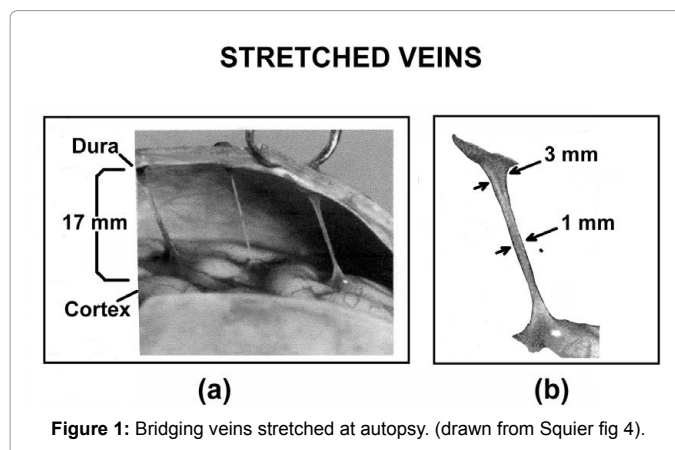


Figure 1: Bridging veins stretched at autopsy. (drawn from Squier fig 4).

subarachnoid space, Sino-Cortical Width (SCW) has been measured with high definition ultrasound by various authors. SCW values in mm are; (1.6+-0.8) [7], (0.4 to 3.3 [8], (3.1 (0.5 to 6 )) [9], (2.8 +-1.33 (1.2 to 5.8)) [10], (3 mm upper limit) [11]. Somewhat less than 3mm seems to be a rough estimate. In Figure 1, the bony strip has been lifted by about 14mm. Although the veins have been stretched, they have not snapped.

There is also a question as to which end would pull out first in vivo if stretching were increased further. The name "Pia" arose from the great care required during dissection to avoid tearing this membrane. Electron microscopy [12] has shown that the Pia Mater itself is only one cell thick, with the cells tightly joined at the edges. Its function is to form a barrier to movement of large molecules. It is not a structural element. Its associated structural components (collagen etc.) lie below, in the subpial space [13]. This delicate physiology suggests that if such tension occurs, it is the Pia Mater (Cortex) end of bridging veins that should tear away before the dural end.

## Experimental Studies of Whiplash Injuries

### Contusions

Contusions are described in ref [14]. Briefly, in 1968, Ommaya et al. [15] secured monkeys on a rail-mounted trolley which could be accurately accelerated by compressed air to simulate collision by a following vehicle. They found that in monkeys, cortical damage was an "All-or-None" phenomenon. Below an estimated 40,000 radians/seconds<sup>2</sup> for 10 msec nothing happened. At higher impact cortical surface hemorrhages, contusions and subarachnoid and subdural hemorrhages appeared. At a certain setting 22 subjects failed to be concussed and 19 were concussed to various degrees. In the non-concussed group not a single animal showed any macroscopic evidence of brain damage. In their table [15] relating to injuries in the concussed group, 4 showed no injuries, and 15 showed injuries. In these 15, contusions (defined as an injury without a break in the skin), were found in 9, (60%). In these whiplash experiments various parts of the brain surface showed evidence of contusions But the AAP definition of SBS says contusions are "unusual" [2]. This implies that it is unusual for current SBS cases to be due to whiplash injuries.

### Summarising Physio-Mechanical Factors Requiring Investigation

This initial survey raised the following points which became the basis of this study of the subarachnoid space.

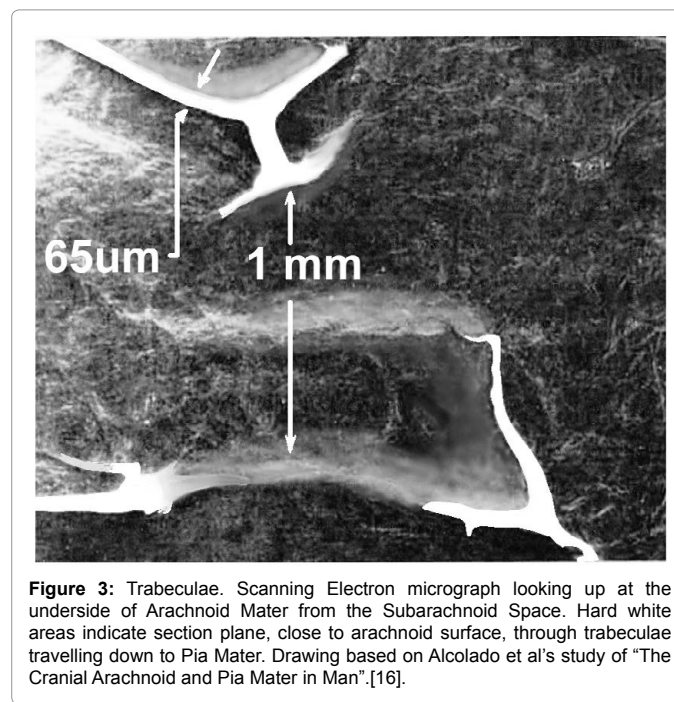
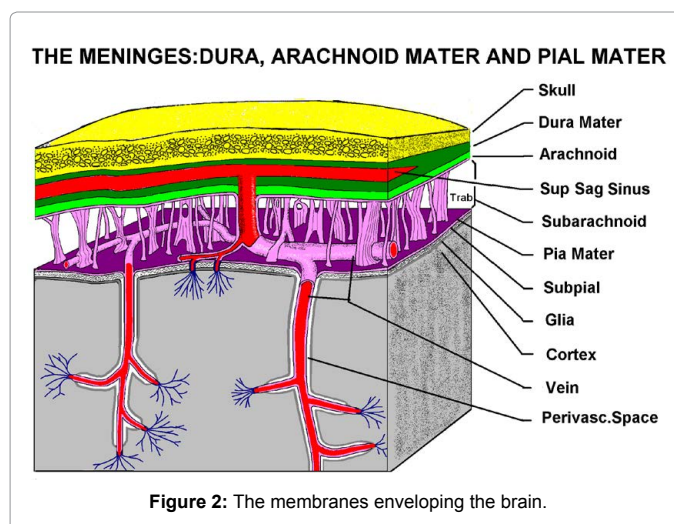
1. Are bridging veins strong enough to cause the damage demonstrated by Guthtech?
2. If they are, which end would pull out first?
3. How much could the brain lag rotation of the skull
4. What type of damage?
5. Veins do not appear to slide in the Dura, Figure 1, how can they cause mechanical damage at the entrance to the superior sagittal sinus?

The first, fundamental and initially surprising finding, was that although the subarachnoid space appears empty on the screens of ultrasound and MRI machines, it is in fact actually filled with a cob-web of collagen reinforced fibres known as "arachnoid trabeculae", Figure 2. "Arachnoid", derived from Greek for spider, refers to the cobweb appearance of this structure. These trabeculae contain fibrocytes and extracellular collagen, and are wrapped by a layer of pial cells [13]. They

are stretched between arachnoid and pia membrane surfaces and define the shape of the subarachnoid space as buttons define the shape of an airbed. They coalesce to form "curtains" Figure 2.

The reason they are not seen on ultrasound machines is that they are extremely thin. MRI scanners can resolve pixels of about 600 μm. The trabeculae in Figure 3 are only about 65 μm thick. Moreover those in Figure 3 occupy less than 1% of the cross section of the picture, so the averaged screen effect would only be a very dark grey, i.e. their presence would not be visible on screen [13]. Similarly, they are too thin to produce echoes on ultrasound and also are well below ultrasound resolution. So on Ultrasound or MRI screens one can see blood vessels crossing between the Pia and Arachnoid membranes but there appears to be nothing else there.

Thus the bridging veins are embedded in a "forest" of very fine trabeculae "curtains", traversing the subarachnoid space, over the cortex and around the spinal cord [16,17]. In the SBS concept, the



bridging veins are completely unsupported, but electron microscopy shows they are attached to this “forest” where ever they pass through or near a curtain. SBS theory says the bridging veins locate the brain relative to the skull. Moore et al. point out that: -“Although it is commonly stated that the brain ‘floats’ in CSF, the brain is suspended in the CSF-filled subarachnoid space by the arachnoid trabeculae” [17]. Thus, the trabeculae bind the brain and skull dura together, profoundly limiting relative movement. The collagen strands within the trabeculae appear to be “stitched” through the inner aspect of the arachnoid at one end and through the pia mater to collagen bundles in the subpial space at the other [16]. The situation appears similar to the structure of clothing. The thread in each stitch is relative weak, but the manufacturer ensures that applied stresses are distributed among sufficient number of stitches to give the garment strength.

The SBS concept appears to have been devised in ignorance of the existence of the trabecular system, or possibly it was assumed that such thin elements could not possibly be strong enough to make any difference. However, while the collagen molecule is slack it is flexible, but when it is stretched, so that its carbon-carbon bonds are lined up, it becomes remarkably strong. Its tensile strength as in catgut is 420 MNm<sup>-2</sup> (Mega Newtons per square metre), compared with Nylon 76-97; Hemp rope 60-100; Aluminium 90-150; and Mild steel (0.2% carbon) 430-490 [18]. Hence its tensile strength is similar to Mild Steel.

The combined traction of hundreds of trabecular micro-curtains ensures that the brain moves with the skull. Arteries and veins are embedded in this trabecular forest, Figure 2, ensure that they move with it, and do not suffer exceptional tension injuries.

### Caffey’s Legacy: Pressure as a Factor

In his ’72 PITS article Caffey noted that Weston had found that subdural hematoma was the primary cause of death. Any alternative hypothesis to SBS has to explain how and why hematomas are formed and why they are first observed in the dura. This requires study of aspects of the dura mater and arachnoid mater which are normally ignored as of little importance and so will be described in detail in the following.

The adjective “forceful”, in the summary at the end of his ’74 paper, is particularly significant. Elsewhere in this paper “forceful” does not appear and vomiting can be read as merely being sick. In the summary he has added “forceful”. Forceful vomiting, sometimes referred to as projectile vomiting, is characteristic of Infantile Hypertrophic Pyloric Stenosis. Taylor, Cass, and Holland describe the current (2012) situation as:- “The aetiology of Infantile Hypertrophic Pyloric Stenosis (IHPS), a condition characterised by abnormal thickening of the pylorus resulting in a gastric outlet obstruction, remains unknown. IHPS typically presents with progressive projectile vomiting that commences between the second and eighth week of life [19,20].

Caffey’s legacy is in this recognition of vomiting in pyloric stenosis as an internal source of pressure. Others had recognised the consequences of excessive venous hypertension but looked in vain for the source of pressure. Guthkelch, like some others, had recognised that raised intrathoracic or intraabdominal pressure would account for the features of “Battered Baby Syndrome”. In his ’71 paper [4] Guthkelch comments on an infant documented to have had several fits of coughing. He says:- *In this case the possibility of compression of the thorax with a consequent rise in jugular venous pressure and rupture of cortical veins is not excluded, but there were no signs of bruising of the chest wall or lungs, nor any rib fractures.* When he could not find evidence of externally applied pressure (bruising etc) he appears to

have dismissed the possibility. He never seems to have considered the possibility of an internal source of hypertension. By drawing attention to forceful vomiting Caffey hinted at pyloric stenosis being involved in some way. In the following the production of Subdural Hematomas is considered, followed by compatibility with other observations from his ’74 paper.

The abdominal contents are bounded by the diaphragm, abdominal muscles, and pelvic floor, Figure 4. Blood from the lower body normally returns to the right atrium of the heart up the Inferior Vena Cava, (IVC). Blood from the cerebrum normally flows down through the jugular veins to the Superior Vena Cava (SVC), where it is joined by blood returning from the arms through the brachiocephalic veins. The combined flow then normally proceeds down the SVC, where it joins blood from the IVC at the entrance to the right atrium of the heart. When high venous pressures, generated in the abdomen by maximal abdominal muscle contractions, arrive at the right atrium, flow in the SVC is reversed. There are valves protecting the brain and eyes, (the inner jugular vein valves), but in humans these are optimised for low forward flow resistance at the expense of withstanding reverse pressure. If the pressure in the SVC rises excessively, the Inner Jugular Vein valves may collapse, allowing blood at high pressure to enter the head veins.

### Retinopathy and Subdural Hematomas

Excessive cerebral venous hypertension is known to produce retinal haemorrhages in a disorder known as Valsalva Retinopathy. Valsalva originally devised his procedure for assessing autonomic (sympathetic) competence [20]. The Valsalva Maneuver involved asking the subject to exhale into a mercury manometer to produce an intrapulmonary pressure of 30 mm Hg, and hold it for a specified time [21-23]. This prevented venous return to the heart, causing peripheral venous pressure to rise. Then the subject was told to breathe normally while his

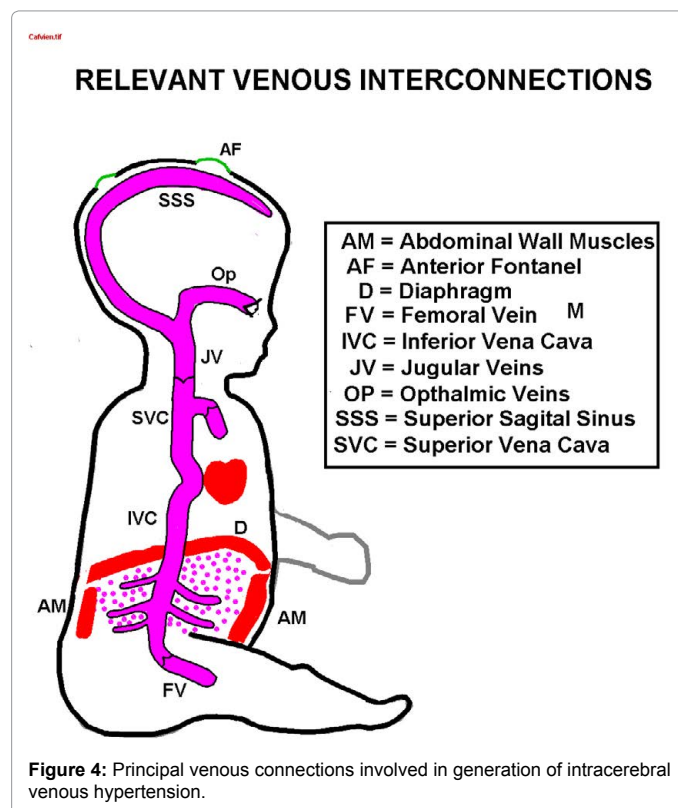
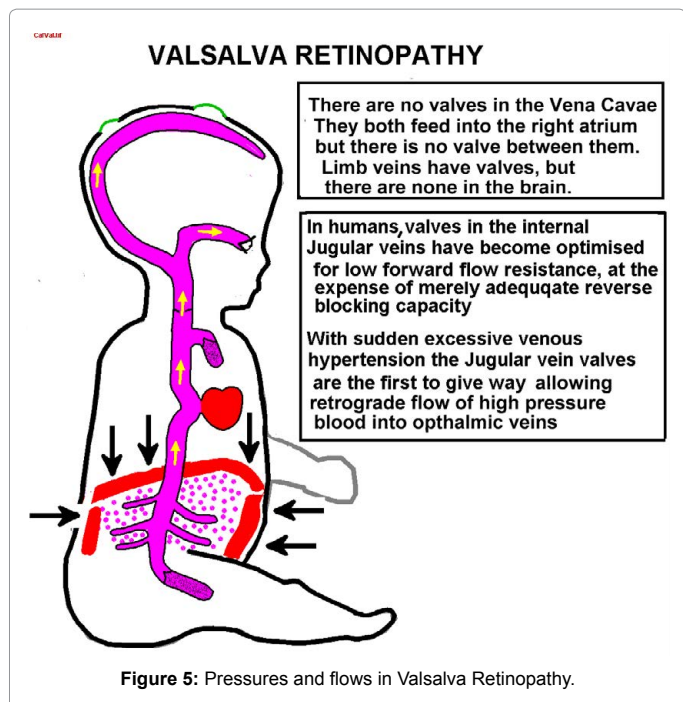


Figure 4: Principal venous connections involved in generation of intracerebral venous hypertension.



arterial pressure was recorded during recovery. The recording showed overshoots etc characteristic of the subject's degree of autonomic control. In 1972, the same year as Caffey's "PITS" paper, Thomas Duane introduced the term "Valsalva Hemorrhagic Retinopathy" to describe the "pathophysiology of retinopathies due to distant trauma". [23]. He recognized that any incident that produced elevated venous pressure would be conveyed throughout the body by the vena caval system and could appear in the ophthalmic vein. He noted that this could result in "small and large transudates and haemorrhages." Duane gives examples of temporary loss of sight, in adults, in blowing up an air mattress, dry heaves (retching, non-productive attempts to vomit), a lady helping her neighbour carry a refrigerator down a flight of stairs, and violent vomiting after a party. All these adults recovered normal vision within a few months.

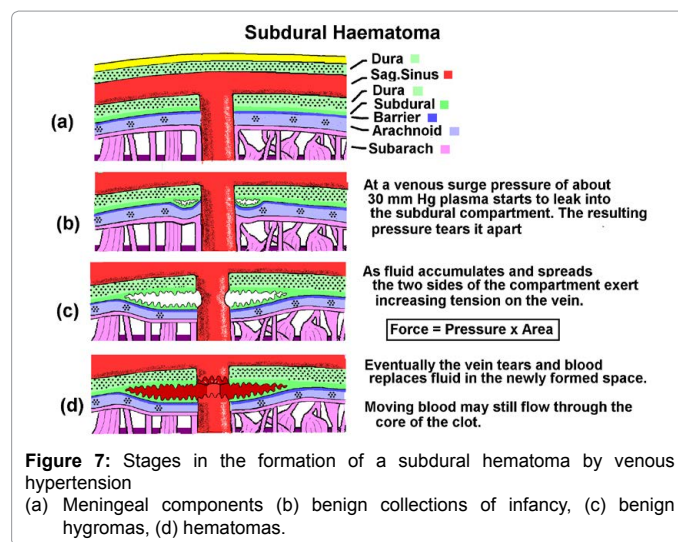
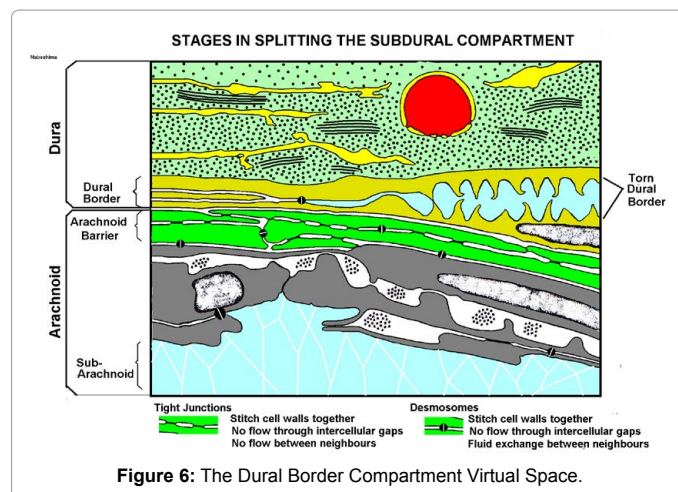
Thus in infants excessive intra-abdominal pressures can be generated by abdominal muscles in violent vomiting or paroxysmal coughing [24]. This pressure will be applied to all blood in the abdomen and will attempt to drive it out of the abdomen, up the IVC, SVC, and into the sagittal sinus and ophthalmic veins, Figure 5. Infantile projectile vomiting may throw the vomit several feet across a room, which demonstrates considerable intra-abdominal pressure. Consequential ophthalmic vein hypertension provides an alternative explanation to shaking in "SBS".

The other common hemorrhagic feature, subdural bleeding, requires more detailed physiological knowledge of the membranes enclosing the brain.

Schachenmayr and Friede [25] showed that the so-called subdural space in man is not a true space, it is a cleavage artifact produced by tearing within the dural border compartment. Orlin et al. [26] studying the dura-arachnoid junction in pigs, described the border layer as consisting of avascular tissue with flake-like relatively electron-lucent cells stacked upon each other in several layers with narrow intercellular clefts. Orlin simulated hemorrhage in this region by fixing a fine cannular at the depth of the subdural compartment, and injecting

autologous blood. The brain was then immediately fixed in situ. They found the compartment split without any particular, internal, cleavage plane. The normal relationship between the dural, arachnoid, and pia maters covering the brain [27] is shown on the left in Figure 6, and after tearing within the subdural compartment on the right. The absence of collagen from the compartment facilitates cleavage by traction from the tougher, collagen reinforced, dura and arachnoid layers. Whereas collagen is extensively present within most of the dura, a physiologically distinct layer exists where it borders on the outer (barrier) layer of the arachnoid. The outer surface of the arachnoid is also specialized. Here cells are tightly joined, forming a barrier to water and solute movement. Unlike this arachnoid barrier layer, cells in the dural border layer are only occasionally bound together. Both layers lack collagen reinforcement, so relative strength depends on cell-to-cell bonding. In this the dural border layer is much weaker than the arachnoid barrier layer, and so it is the dural border compartment that gets torn apart.

Yamashima and Friede studied bridging veins by electron microscopy [28] and found that whereas in the subarachnoid portion vein walls were well supported with collagen, where they passed through the Dura thickness varied widely, wall thickness could be as little as 10 μm. There was a tendency for more circumferential than longitudinal collagen fibres (Figure 7a). They suggested that "Conceivably, a physiological increase of intravenous pressure, for example



from defaecation or coughing could cause leakage at the weakest portions of the bridging veins, even in the absence of trauma. Friede commented that: - thin, fresh subdural haemorrhages found at necropsy often correlate with terminal external cardiac massage causing violent compression of the thorax or even rib fractures. .... In conclusion one may state that not only sudden acceleration or deceleration of the head but also sudden increases in venous pressure can lead to an augmentation of tension, especially at the subdural portion of bridging veins, thus inducing subdural bleeding.

The pressure surge during vomiting is one such sudden increase in venous pressure. When a pressure surge arrives in the sagittal sinus the bridging vein will leak water at this point (Figure 7b).

Even if a pressure surge is insufficient to damage large veins, local dural venules may leak. Mayhan and Heistad [29] raised pial vein pressure in rats from  $7 \pm 1$  mmHg to  $30 \pm 3$  mmHg by occluding their superior venae cavae. They detected leakage with fluorescein labeled dextran, (mol wt. 70,000) and found it normally leaked at  $0.002 \pm 0.001$   $\mu$ l/sec but rose to  $0.31 \pm 0.059$   $\mu$ l/sec at 30 mmHg. This increase then lasted for at least 40 minutes after venous pressure had been returned to normal, indicating some form of structural damage.

This subdural region is unique in that extravasated water lies between the impervious skull and the arachnoid barrier layer. It cannot drain easily, resulting in a form of edema which will push the surfaces of the subdural compartment apart. This region of the bridging vein is attached to both sides of the subdural compartment. As the sides are forced apart they put tension on this section of the vein. This tension increases as the edemous pressure is applied to an increasing area as the subdural compartment starts to tear apart. (Force=Pressure  $\times$  area) (Figure 7c).

The space between the surfaces increasingly fills with fluid (Hygroma). Eventually the vein wall tears, Figure 7d. Blood then fills this hygroma space and venous hypertensive surges can act directly to further split the subdural compartment, forming a subdural hematoma. There is nowhere else along a bridging vein where extravasated water can build up tension and rupture them which is why hematomas appear first at this subdural site.

### The Pyloric Mechanism and Male Infant Preponderance in SBS Convictions

Miller and Miller studied the gender ratio of the incidence of TBII (Traumatic Brain Injuries of Infancy) attributed to various situations [30]. Relevant scientific articles published between 1966 and 2005 were found using the keywords “Shaken Baby Syndrome”, “Retinal Hemorrhage”, and “Subdural Hematoma” to search using Medline. In infants with subdural haemorrhages who were the subject of Shaken Baby Syndrome convictions 62.6% were male, a substantial male predominance. This 2:1 male predominance is hard to explain in terms of shaking but arises naturally in the pyloric stenosis hypothesis [31].

The mechanism of pyloric stenosis is dealt with in some detail in references [32,33]. Briefly, the pylorus is a section of the gut between the stomach and intestine, Figure 8. Its main components are the Pyloric Sphincter, (which operates open or shut) and the Pyloric Canal which operates independently, but cooperatively, as a pump. The pylorus is not just a tap to regulate transmission of partially digested food from the stomach to the intestine; it is an organ in its own right. With the pyloric sphincter remaining closed the pyloric canal muscle continually squirts partially digested food back into the stomach antrum. The resulting viscous forces strip off the softened outer layer of food lumps returning them to the stomach for further chemical reduction. This is sometimes

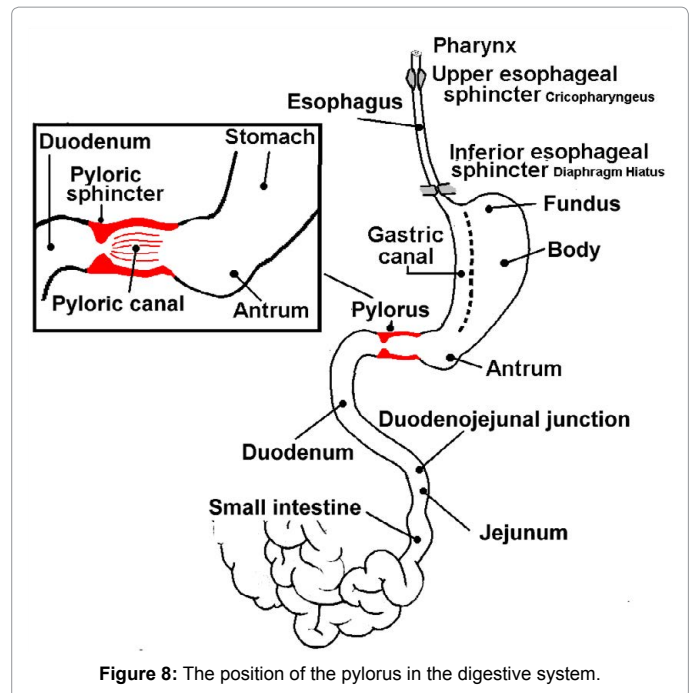
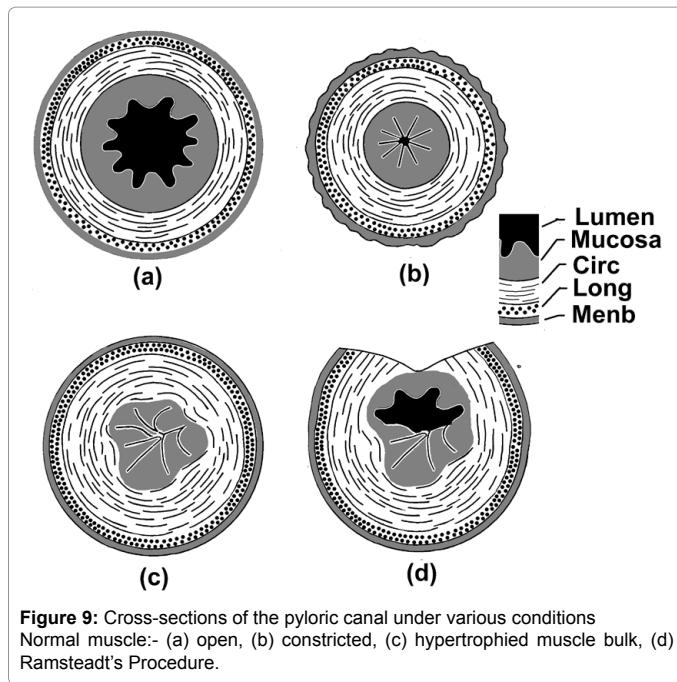


Figure 8: The position of the pylorus in the digestive system.

referred to as “grinding” but that is misleading. Birds and reptiles have a grinding apparatus with hardened surfaces, (the Gizzard) [34]. The mammalian viscous stripping method requires sudden powerful contractions of the pyloric canal muscle.

The pylorus forms late in fetal life and at birth is very immature [35,36] though adequate for a liquid diet. Muscle bulk needs to increase during the first few months of postnatal life ready for weaning. Challa et al. [37], in their EM study of infants with hypertrophic pyloric stenosis [37], could find no ultrastructural abnormalities in the muscle cells. In the neuropil, swollen axons were observed which they attributed to mechanical compression by the hypertrophied muscle. So what goes wrong with this process to result in hypertrophic stenosis? The answer appears to lie in the stress transmission features of the muscle cells themselves.

Stomach and intestinal smooth muscle cells are extremely elongated, typically 0.2 mm (200  $\mu$ m) long and up to 8  $\mu$ m wide; i.e. a length to width ratio of 25:1. Most (80-90%) of the cell volume is packed with actin-myosin contractile filaments [31]. Each smooth muscle cell is externally bounded by a basement membrane. Contiguous visceral smooth muscle cells communicate electrically through gap junctions similar to those between cardiac muscle cells [38], enabling them to cooperate as unitary muscles. They can operate independently of innervation, spontaneously setting their own rhythmic contraction pattern. The function of innervation in visceral muscles is to modulate that pattern, speeding it up or slowing it down, strengthening or weakening it. Like voluntary muscle, visceral muscle uses an actin-myosin “motor” system to power contraction, but in smooth muscle there are many more actin filaments than myosin, 2:1 in voluntary and 12:1 in smooth [39]. They are not neatly arranged as in voluntary muscle. In smooth muscle myosin occurs in long ribbons surrounded by many actin filaments. This system enables smooth muscles to have a greater range of contraction, (4:1), than voluntary muscle, [40] but they need protection against overextension. This is provided by inextensible filaments of desmin around and within individual cells, and collagen and reticulin fibres between cells. These form a “safety net”. Cells can



**Figure 9:** Cross-sections of the pyloric canal under various conditions  
Normal muscle:- (a) open, (b) constricted, (c) hypertrophied muscle bulk, (d) Ramsteadt's Procedure.

be stretched freely until these safety nets are pulled tight, then they strongly resist further extension.

### Stenosis and the Influence of Testosterone

Cross sections of the pyloric canal in various conditions are shown in Figure 9. In (Figure 9a) and (Figure 9b) the muscle is normal and functional. In (Figure 9a) the circular muscle is relaxed and the lumen is open with longitudinal folds in the mucosa. In (b) the circular muscle has contracted. It looks thicker because the same muscle mass is crowded into a smaller space. Similarly the longitudinal layer has been pulled in and the outer layer has puckered. In (9c) production of contractile proteins within smooth muscle cells has been accelerated. The muscle cell volumes have increased, but the circumference of the circular muscle cannot increase in proportion because it's cell "safety nets" are stretched tight. The greater muscle volume can only be accommodated at the expense of the lumen volume, leading to occlusion. This has to be surgically relieved, usually by Ramsteadt's procedure, Figure 9d [41]. A cut is made along the pyloric canal to a depth just reaching the mucosa (Figure 9d). This relieves the tension in the circular muscle and the lumen can then open.

Guthkelch [42], commenting on the incidence of subdural haematoma in his 4-yr series of infants admitted for fits and/or vomiting, quoted Ingraham and Matson as finding a preponderance of males over females in the ratio of 5:3, and in his own series a preponderance of 5:1. Other workers have reported similar M/F gender ratios 4:1 M/F [43], 5:1 [17], 4:1 to 6:1 [44] 5.6:1 [19]. Males also predominate by 2:1 in infants the subject of SBS convictions [30].

An explanation of this bias is given in reference [31]. In brief, Testosterone accelerates the production of contractile proteins within cells, i.e. it increases muscle bulk, but not maximum stretched length. (Sports drug cheats increase muscle bulk with testosterone but their muscles do not get longer and floppy). Hence for the same rate of cell production males would be expected to reach pyloric stenosis earlier than females, producing male the preponderance.

### Conclusions

The AAP 1993 definition of SBS [2] does not take into account the presence and structural function of arachnoid trabeculae. It is this "cob-web" of collagen reinforced trabeculae, not the bridging veins, that takes the strain of rotational movement of the skull on the brain and vessels in the subarachnoid space. Moreover, this 1993 SBS definition reports that visible cerebral contusions are unusual, but that subdural hemorrhage is common. This is the reverse of the experimental findings of Ommaya et al. [15] and Gennaralli [45]. Hence the current definition of SBS is based on a fallacious model, and it does not match experimental behavior. It is invalid, as are any convictions based upon it.

However, the AAP listing of clinical features preceding fatal events is appropriate and valuable. *The infant may have a history of poor feeding, vomiting, lethargy, and/or irritability occurring for days or weeks prior to the time of initial health care contact. The subtle symptoms are often minimized by physicians or attributed to mild viral illnesses, feeding dysfunction, or infant colic.* These features should alert one to the possibility of pyloric stenosis, which can readily be investigated ultrasonically, or, if at an advanced stage, the pylorus can be felt as an "olive" under the skin. Not mentioned in this list are the bouts of inconsolable crying which were suggested to stress the carer to the point of violent shaking. Forceful retching or vomiting can lead to tears at the junction of the oesophagus and stomach, Mallory-Weiss tears [32]. These infants are not just irritable, they are in genuine pain.

Caffey's legacy can be summed up, with one word, ("Forceful"), in one sentence, at the end of his last publication on SBS. Others had suspected vascular hypertension could be involved but had sought its origin in externally applied trauma. Caffey provided an internal source of pressure. Though he continued to believe that the injuries were caused mechanically by shaking, he recorded an apparently unconnected association with bulging fontanels and projectile vomiting. His reputation for an honest scientific approach to medicine is illustrated by his willingness to publish these associations, which could potentially provide an alternative to his shaking hypothesis.

Summarising the new hypothesis,

1. The hypothesis suggested here is that transient excessive abdominal pressures generated during vomiting or retching arrive in the veins of the eyes and brain and cause the hemorrhages.
2. Subdural haematomas arise when venous hypertension surges force fluid through the vein walls into the weak subdural compartment and split it.
3. Since pyloric muscle development takes place in the first few post-natal months it is understandable that this disorder should peak at this time.
4. Male predominance in pyloric stenosis is due to accelerated contractile protein production in pylorus muscle cells caused by the action of testosterone. The predominance of male infants in SBS convictions reflects this male predominance in the causative pyloric stenosis.
5. This alternative pressure mechanism has previously been described as Dysphagic Infant Death Syndrome (DIDS) [32]. DIDS does not involve head movement; it is purely hydromechanical and can occur while the head is static. It is physiological and does not involve the carer, who is innocent of imposed trauma.

It has been said of Dr Caffey that he “*was full of paradox and contradiction. He valued the life of the intellect highly, and he was well aware that he had been particularly blessed in mental acuity. He did not suffer fools gladly.*” Dr Caffey was said to be “*a scholar in the best sense of the word; he loved to dig to the bottom of whatever was being considered, and he was devoted to the truth. He was sceptical of conventional explanations and was in the habit of disbelieving other authorities.*”

He published his 1974 paper after he had retired. He was still working daily, and at weekends, in an emeritus position in the Childrens Hospital of Pittsburg, at the age of 79, just four years before his death in 1978.

Unfortunately he died (at 83) before he could have followed up this “gut feeling”, but he left the clues for others to follow up.

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