

# MAJOR REVIEW

## Fundus Hemorrhages in Infancy

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**Abstract.** The anatomical location and appearance of retinal hemorrhages in the infant provide important clues in the diagnosis of underlying disorders. While neonatal retinal hemorrhages related to birth trauma are common, benign, and self-limited, other retinal hemorrhages in infancy may signify intracranial aneurysms, accidental or non-accidental injury, and a variety of ocular (e.g., Coats' disease, PHPV, ROP, retinal dysplasia, hypertension, myopia) or systemic disease (e.g., hematologic or cardiovascular disorders, infection, protein C deficiency). In this review, retinal hemorrhages are illustrated and classified according to location, appearance, and etiology. Prompt diagnosis of retinal hemorrhages in infants is crucial, because treatment may be required to prevent early deprivation amblyopia and blindness. Ophthalmological findings may also be a valuable contribution to the overall medical evaluation of the infant. (*Surv Ophthalmol* 37:1-17, 1992)

**Key words.** central venous pressure • child abuse • infantile retinal hemorrhages • intracranial pressure • intraocular venous pressure • non-accidental injury • retinal hemorrhages • subarachnoid hemorrhage

Retinal hemorrhages, although nonspecific, may be the presenting sign of an underlying disease or disorder in the infant. Recognition of retinal hemorrhages and other associated signs guide the physician to appropriate investigation and management of the hemorrhages, as well as the underlying disorder. While some hemorrhages resolve spontaneously, others require prompt treatment to prevent early deprivation amblyopia and blindness.

### I. Clinical Appearance

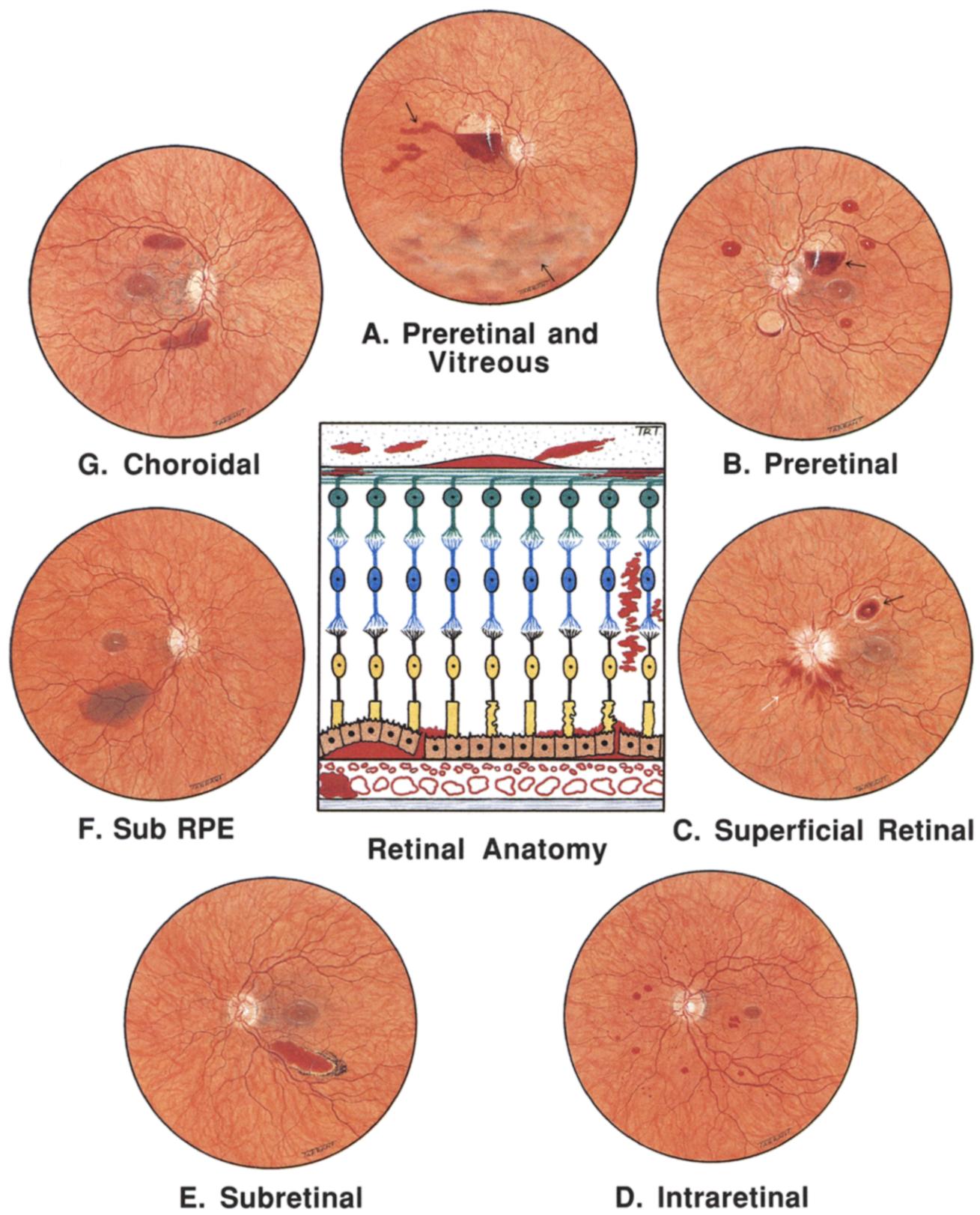
Retinal hemorrhages occur when there is extravasation of blood, usually from capillaries or postcapillary venules. More severe hemorrhages result from diseased larger venules or arterioles. The clinical appearance of retinal hemorrhages varies according to the site of the extravasation of the blood. Retinal hemorrhages may involve all the retinal layers or they may be confined to one layer only. They may be unilateral or bilateral, localized or generalized. They are usually most numerous centrally where the retina is thickest and where the capillary circulation is more com-

plex than in the periphery. Richman in 1936 classified retinal hemorrhages into four groups according to their morphological appearances<sup>89</sup>: today, clinically, they can be divided into intravitrreal, preretinal, intraretinal, subretinal, subretinal pigment epithelial and choroidal hemorrhages (Fig. 1).

### II. Pathological Anatomy

#### A. VITREOUS HEMORRHAGES

Larger preretinal and intraretinal hemorrhages may penetrate into the normally avascular vitreous gel and appear as curls or linear streaks in formed vitreous or as diffuse hemorrhage in fluid vitreous (Fig. 1A). Because of the very compact gel nature of infant vitreous, intra-gel hemorrhages usually take a long time to clear and they may become loculated. Unless they are very severe, they usually clear completely,<sup>5</sup> but leave an abnormal vitreous structure. Recurrent hemorrhages delay clearing of the vitreous and altered blood may become compacted in the posterior gel to form an "ochre membrane." In pa-



**Fig. 1. Anatomical Description of Fundus Hemorrhages**

*Fig. 1.* Centerpiece: Cross-sectional drawing of the retina showing the various sites of hemorrhages as detailed in the fundus view paintings A-G. *A:* Preretinal hemorrhage with small vitreous hemorrhage (arrows). *B:* Preretinal hemorrhage (arrow). The blood strips up the internal limiting membrane or the hyaloid face of the vitreous body and is thus contained, often with a "feathered" edge. The small hemorrhages are intraretinal, the central white spot being a light reflex on the domed surface. *C:* Superficial intraretinal hemorrhages (white arrow). The flame-shaped hemorrhages originate from the superficial peripapillary capillaries. The larger hemorrhage (black arrow) is intraretinal with breakout into the subhyaloid space. *D:* Deep intraretinal hemorrhages. The larger "blot" hemorrhages are full thickness and often longstanding. The smaller "dot" hemorrhages are round and uniform and disappear without a trace. *E:* Subretinal hemorrhage. These occur between photoreceptors and the retinal pigment epithelium. As they absorb, they usually leave a pigmented area. They are associated with a dense scotoma. *F:* Sub-RPE hemorrhage. Hemorrhages beneath the RPE usually represent a developing disciform lesion. The RPE is smoothly elevated and slate-red in color. *G:* Choroidal hemorrhages. These are usually traumatic and may not disturb the overlying retina. They may be difficult to see, appearing as an often extensive smooth elevation of the retina, being homogenous as compared with the normal choroid pattern. (Artist: Mr. Terry Tarrant, London, England.)

tients with retinal neovascularization, as in retinopathy of prematurity, vitreous hemorrhage is a frequent cause of complications.

Vitreous hemorrhages are uncommon in infancy,<sup>5</sup> and early vitrectomy may be necessary to prevent deprivation amblyopia. In our experience, the vitrectomy must be followed by occlusion of the uninvolved eye in unilateral cases.

## B. PRERETINAL HEMORRHAGES

Preretinal hemorrhages, first described by Liebreich (1863),<sup>62</sup> result from bleeding into the subhyaloid space between the internal limiting membrane of the retina and the posterior vitreous face (Fig. 1B). We believe that the typical

preretinal hemorrhage is 1–2 disc diameters (dd) in size, near the posterior pole, often at the macula. Hemorrhages are often multiple and extensive. The blood spreads on the retinal surface as a dark red mass; it is round and densest centrally when fresh. It may settle with gravity, forming a horizontal fluid level [hence, the term, "boat-shaped" (Fig. 2) or crescentic hemorrhage]. The lower curved portion is dark red, and the color fades to a yellow-white toward the horizontal line above. This plasma-erythrocyte interface shifts with changes in the position of the head.<sup>69</sup> The absorption usually occurs from above, because the upper layers are lighter and less dense than the lower, which absorb and disappear last and may leave a delicate curved line representing the lower border (Fig. 2).

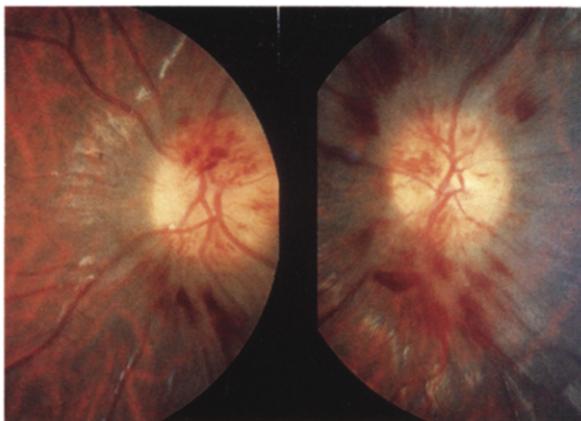
"Thumbprint"<sup>40</sup> preretinal hemorrhages occur at the posterior pole; they usually number 3–8, and each is about one dd in size. They are darkest in the center and show "fraying" in their



*Fig. 2.* A boat-shaped preretinal hemorrhage. In this instance, the boat-shaped hemorrhage, which has a horizontal fluid level, probably is contained by the internal limiting membrane of the retina at the macula, while there is additional fluid between the internal limiting membrane and the hyaloid layer of the vitreous. The infant had a bleeding diathesis associated with treated leukemia.



*Fig. 3.* Thumbprint preretinal hemorrhage in a patient with leukemia.



*Fig. 4.* This infant with neuroblastoma had an acute optic neuropathy; the optic discs show multiple flame-shaped hemorrhages, pallor and swelling.



*Fig. 5.* The retina of this child with aplastic anemia shows a few mid-peripheral flame-shaped hemorrhages from bleeding into the nerve fiber layer.

extremities (Fig. 3). Ophthalmoscopically, there is a small, glistening central light reflex, which moves with changes in the viewing angle.

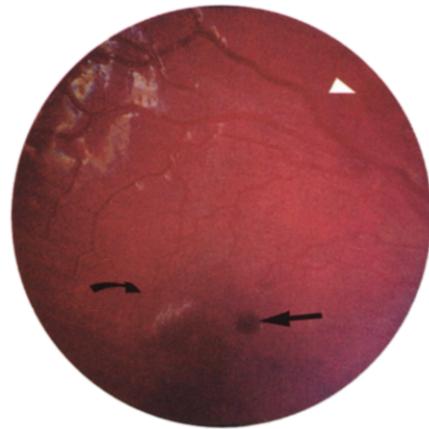
Posterior polar preretinal hemorrhages are common in children with either subdural or subarachnoid bleeding.<sup>41</sup> These hemorrhages may be facilitated by the relatively thin internal limiting membrane in the infant.<sup>27</sup>

Clinically, the hemorrhage may appear to lie between the retina and the vitreous.<sup>53,76</sup> However, some pathological studies have shown that it lies between the nerve fiber layer and the internal limiting membrane,<sup>28</sup> while others depict it between the internal limiting membrane and the hyaloid layer of the vitreous body.<sup>108</sup>

### C. INTRARETINAL HEMORRHAGES

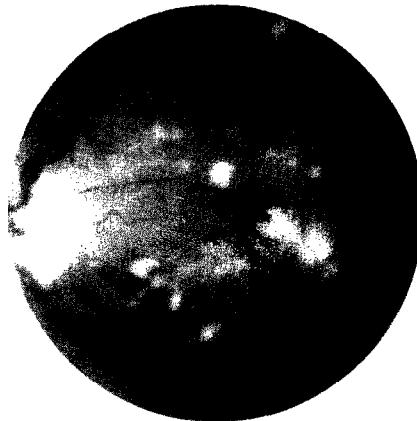
Superficial retinal hemorrhages are "splinter" or "flame," and originate from the superficial capillary bed or the superficial radial peripapillary capillaries (Fig. 1C). Blood pools in the direction of least resistance among the nerve fibers; thus, they appear bright red and elongated with frayed distal borders in the distribution of the nerve fibers. They occur especially around the optic disc (Fig. 4). They are seldom seen in the mid- (Fig. 5) or peripheral retina,<sup>7</sup> because about 7 mm from the disc the nerve fiber bundles form a more open network with round or polygonal interstices,<sup>7</sup> causing even superficial hemorrhages to assume an irregular rounded form.<sup>74</sup> They usually clear rapidly, even within a few days.<sup>101</sup> These hemorrhages suggest disease affecting the superficial peripapillary capillaries.<sup>40</sup>

Deep retinal hemorrhages, often called "dot and blot" (Fig. 1D), are located in the inner nu-

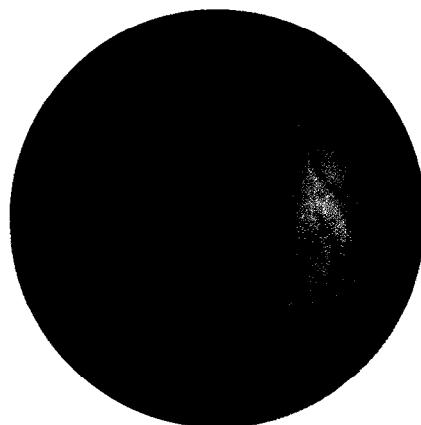


*Fig. 6.* A blot hemorrhage (black arrow) near the fovea consists of a full-thickness collection of blood. Flame-shaped hemorrhages (white arrow) are also present in the peripapillary nerve fiber layer, and there is a small subretinal hemorrhage (curved arrow). The child has thrombocytopenia.

clear layer and spread to the outer plexiform layer.<sup>119</sup> The vertical arrangements of Müller's fibers and neurones limit lateral spread. In the peripheral retina, the nerve fiber layer is thin, so most retinal hemorrhages are dot and blot type. They usually occur with disorders affecting the deep retinal capillaries. A dot hemorrhage is a small cluster of red cells and may be confused ophthalmoscopically with a retinal microaneurysm. It is round and of uniform density, and disappears without trace.<sup>40</sup> Blot hemorrhages are full-thickness retinal hemorrhages (Fig. 6), which are often a feature of venous occlusion and suggest retinal ischemia (Fig. 7). They are dark



*Fig. 7.* Intraretinal hemorrhages and cotton wool spots in a child with cytomegalovirus retinitis. He was in remission for acute lymphoblastic leukemia.



*Fig. 8.* A blotchy red subretinal hemorrhage lies between the photoreceptors and the pigment epithelium.

red, rounded, and in a three-dimensional view, they appear domed.<sup>119</sup>

#### D. SUBRETINAL HEMORRHAGES

Subretinal hemorrhages appear as a blotchy red areas, with the retina elevated (Fig. 1E). They occur between the photoreceptors and pigment epithelium (Fig. 8). Macular hemorrhages are frequently associated with choroidal subretinal neovascularization or trauma. As they absorb they can become depigmented, giving a whitish-yellow appearance. Mechanical interference with the choriocapillaries account for the variable permanent central scotoma. The hemorrhages may extend from large, deep retinal hemorrhages that break through the external limiting membrane, as in Coats' disease,<sup>13,90</sup> sickle cell disease,<sup>105</sup> leukemia,<sup>93</sup> retinopathy of prematurity,<sup>106</sup> and angiomas retinae.<sup>40</sup> They may also result from ruptured subretinal new vessels, as in Coats' disease.

#### E. SUBRETINAL PIGMENT EPITHELIAL HEMORRHAGES (SUB-RPE)

Hemorrhages beneath the pigment epithelium (hematoma of the pigment epithelium,<sup>33</sup> hemorrhagic detachment of the pigment epithelium) are derived from the choroid. They usually represent a developing disciform macular lesion. They enter the space between Bruch's membrane and the retinal pigment epithelium through a break in the Bruch's membrane or at the disc border. The pigment epithelium is usually elevated, with smooth, rounded borders and a dark slate color (Fig. 1F). The RPE stretches as the mound develops, and commonly breaks at

the edge, producing a pathognomonic red corona of subretinal blood.<sup>40</sup>

#### F. CHOROIDAL HEMORRHAGES

Choroidal hemorrhages are dark reddish and often extensive. They are usually traumatic and occur at the posterior pole (Fig. 1G). They may appear with little overlying retinal pathology and usually absorb, leaving a normal fundus.

#### G. WHITE CENTERED HEMORRHAGES

These white-centered retinal hemorrhages were first described by Litten,<sup>63</sup> who gave them the name "Roth spots." Roth,<sup>94</sup> in his original publication in 1872, described white spots as well as hemorrhages. However, he made no association between the two and did not relate these spots to endocarditis. Roth spots are hemorrhages with a pale white center (Figs. 9 and 10), which can be focal ischemia, an inflammatory infiltrate, a colony of infectious organisms, fibrin and platelets, or an accumulation of neoplastic cells.<sup>29,54</sup> They often absorb, leaving no trace.

### III. Associated Conditions and Etiology

#### A. NEONATAL HEMORRHAGES

Retinal hemorrhages in the newborn, first recorded and illustrated by von Jäger (1861),<sup>45</sup> are a benign form of hemorrhagic retinopathy, which occur in the neonatal period. Many studies have been concerned with the incidence, morphology, etiology and significance of these

TABLE 1  
*Types and Main Features of Fundus Hemorrhages in Infancy*

Condition	Frequency of Hemorrhages in the Condition	Shape and Site in the Fundus	Associated Ocular Findings	Associated Systemic Findings	Resorption and Ocular Sequelae
Neonatal retinal hemorrhages	30–40% within 1 hour of birth <sup>33,99</sup>	Splinter-shaped, flame-shaped & dot & blot <sup>8,114</sup>	Subconjunctival hemorrhage	None	Resorption nearly always total and rapid <sup>8,101</sup>
Child abuse	65–89% <sup>4,11,30</sup>	All retinal layers, different "ages" & stages of resorption in different areas. Vitreous hemorrhages <sup>48,60,81</sup> frequent	Retinoschisis, retinal detachment, dislocated lenses. Severity of intracranial hemorrhage correlates with severity of intraocular hemorrhage	Widespread bruises, fractures & intracerebral hemorrhage.	Prolonged resorption, may take weeks. <sup>48,60</sup> Residual scarring & optic atrophy <sup>36,37</sup>
Fundus hemorrhages with subarachnoid hemorrhages	20% <sup>77</sup>	Retinal or preretinal. Occasionally vitreous <sup>11</sup> usually around the disc	Papilloedema	Subarachnoid hemorrhage	Small hemorrhages reabsorb well; large preretinal or vitreous hemorrhages may require vitrectomy to prevent amblyopia in an infant.
Leukemia	High	All layers. White centered hemorrhages, <sup>21</sup> Preretinal & vitreous hemorrhage frequent	Infiltration of all other areas, especially uvea. Retinal infarcts <sup>56,121</sup>	Systemic features of leukemia	Good, given induction of remission
Other blood dyscrasias	Moderate	All layers of retinal & vitreous hemorrhage	Hemorrhages in other tissues	Bruising, bleeding	Usually resorb fully, unless extensive
Hyperviscosity	Moderate	All retinal layers, dot & blot & flame-shaped	Uveal vascular engorgement. Retinal vessel tortuosity	Dark blue skin hue, edema. Cystic fibrosis	Prognosis usually good
C-C fistula	C-C fistula is rare in infancy	Peripapillary & vitreous	Proptosis, papilledema, vascular engorgement, glaucoma	Bruit, traumatic skull lesions, cranial nerve palsies	Prognosis good, given spontaneous or induced remission
PHPV	Low	Intraretinal & vitreous hemorrhages <sup>52</sup>	Microphthalmos cataract with intralenticular hemorrhage, retrolental membrane	None	Poor

TABLE 1  
*Types and Main Features of Fundus Hemorrhages in Infancy (Continued)*

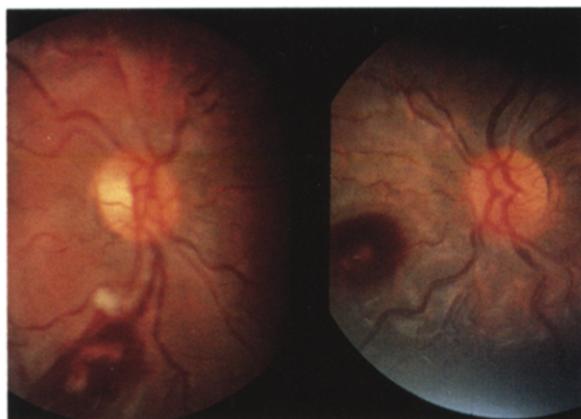
Condition	Frequency of Hemorrhages in the Condition	Shape and Site in the Fundus	Associated Ocular Findings	Associated Systemic Findings	Resorption and Ocular Sequelae
Coats' disease	Moderate	Intraretinal & vitreous <sup>87</sup>	Subretinal exudate, telangiectatic vessels	None	Poor, with residual damage even in cases with benign outcome
ROP	High in progressive disease	Intraretinal, usually near neovascular ridge. Vitreous <sup>97</sup>	Retinal new vessels, ridge 'plus' disease	Prematurity	Moderate to high incidence of sequelae, mainly due to associated disease
Retinal dysplasia	High	Blood lakes in dysplastic retina	Retinal dysplasia shallow AC, glaucoma	None, except in Norries' disease	Very poor prognosis
Hypertension	High	Peripapillary, flame-shaped, occasionally vitreous	Papilledema, retinal exudates, infarcts, choroidal infarcts	Hypertensive encephalopathy. Renal disease	Few sequelae, if there is careful and timely blood pressure control
Myopia	Low	Intraretinal, subretinal with disciform lesions, vitreous	Chorioretinal disease in progressive myopic detachment	None, unless associated with systemic condition i.e. Stickler's, Kniest's or Marfan's syndrome	Good, except with disciform lesion
Syndrome of dominant tortuosity with retinal hemorrhages	Uncertain	Superficial & preretinal	Retinal arterial tortuosity, especially around macula	None	Spontaneous resolution <sup>117</sup>
Retinal infections	High	Small intraretinal hemorrhages in all layers in areas of retinal necrosis <sup>100</sup>	Retinal necrosis, cotton wool spots, "crumbled cheese & ketchup"	Systemic signs of infection and/or immune deficiency	

This table includes only the most important characteristics; see text for additional information. Table combines authors' personal observations with literature reports.

hemorrhages.<sup>8,10,20,32,45,47,52,57,68,88,98,100,113</sup> Their reported incidence has varied between 2.6% and 59%.<sup>8,10,19,20,33,43,46,53,69,71,89,99,101,114</sup> These variations have been influenced by the age at ophthalmological examination, the type of delivery, the parity of the mother, the ophthalmoscopic tech-

nique used, and the examiner (i.e., experienced or inexperienced ophthalmologist, a pediatrician, or other).<sup>114</sup>

Within one hour of birth, the incidence is about 40%, but it decreases to 11% by 72 hours.<sup>33,99</sup> Retinal hemorrhages occur in about



*Fig. 9.* White centered hemorrhages in an infant with acute lymphoblastic leukemia in relapse.



*Fig. 10.* White centered hemorrhage in a child with acute lymphoblastic leukemia.

one-third of babies born by occipital presentation or induced labor, and the incidence increases when obstetric procedures are used, in prolonged labor,<sup>5,53</sup> when the mother is toxicemic,<sup>102</sup> and in older primiparae. It is significantly lower in cesarean section and breech deliveries.<sup>114</sup> The frequency of retinal hemorrhages in induced labor may depend on the inducing agent used.<sup>99</sup> Schoenfeld et al<sup>99</sup> reported that the incidence was 28% in oxytocin-induced deliveries and 40% in dinoprostone-induced deliveries; the difference was attributed to the prostaglandin properties of dinoprostone. Retinal hemorrhages have been found to be more frequent in premature infants treated with parenteral prophylactic vitamin E.<sup>92</sup> Bilateral vitreous hemorrhage has been recorded,<sup>78</sup> and has also been found at birth in protein C deficiency.<sup>86</sup>

Neonatal retinal hemorrhages may be caused by compression of the head within the birth canal, thereby increasing intracranial pressure, unequal pressure distribution during birth,<sup>20,98</sup> prostaglandins in the fetal circulation,<sup>99</sup> and altered blood viscosity.<sup>8</sup> Release of endogenous prostaglandins after amniotomy or in oxytocin treatment may transiently break down the blood retinal barrier.<sup>99</sup> After cesarean section, there is no release of prostaglandins, which combined with the less marked pressure changes is perhaps why retinal hemorrhages in these neonates is rare.<sup>61,98</sup> Neonatal retinal hemorrhages are usually a mixture of splinter-shaped, flame-shaped and dot and blot hemorrhages,<sup>8,114</sup> with rare subretinal and preretinal hemorrhages (Fig. 11). They are usually at the posterior pole and resolve within a few days.<sup>8,101</sup> Macular hemor-

rhages have been found in about 4% of births<sup>43</sup>; these, too, usually resolve completely, and follow-up studies have reported normal visual development.<sup>44,95</sup> Prolonged macular hemorrhage, however, may cause deprivation amblyopia.<sup>43</sup> Vitrectomy may need to be considered in early onset severe vitreous hemorrhage to prevent amblyopia.

#### B. SUBARACHNOID AND SUBDURAL HEMORRHAGES

Subarachnoid hemorrhage results when bleeding occurs into the subarachnoid space following ruptured intracranial aneurysms<sup>66,112</sup> or after craniocerebral trauma.<sup>111</sup> Intraocular hemorrhages occur in about 20%<sup>77</sup> of patients with subarachnoid hemorrhage; they occur simultaneously or are seen within a few days.<sup>25,67</sup> The mortality rate of subarachnoid hemorrhage is about 50%<sup>77</sup> when intraocular hemorrhages are present, but only 25% when they are absent, and it is higher in bilateral than in unilateral hemorrhages.<sup>111</sup>

The intraocular hemorrhages are usually retinal or preretinal (Figs. 1B, 2, and 3). They are at or near the optic disc, occasionally breaking into the vitreous.<sup>111</sup> They were first described by Tersteson in 1912,<sup>107</sup> but the pathogenesis of these intraocular hemorrhages and the associated optic nerve sheath hemorrhages is controversial. Ballantyne in 1943<sup>6</sup> thought that the intraocular hemorrhage and optic nerve sheath hemorrhage both resulted from a sudden rise in intracranial pressure causing stasis in veins draining from the eye and orbit. However, Muller and Deck<sup>77</sup> showed that cavernous sinus flow was not ob-

structed even up to an intracranial pressure of 250 mm Hg. The ophthalmic vessels drain into the cavernous sinus, and the facial and pterygoid veins and the central retinal vein have a major connection with the superior ophthalmic vein. Hence, any venous obstruction at the level of cavernous sinus would not substantially affect the central retinal vein.

Another view<sup>83,104</sup> was that the subarachnoid blood is forced into the optic nerve sheath and penetrates the lamina cribrosa to appear within the globe. Intraocular and optic nerve sheath hemorrhages have been reported in cases of sudden intracranial bleeding.<sup>115</sup> Muller and Deck<sup>77</sup> observed optic nerve sheath hemorrhage in a patient with cerebral swelling without subarachnoid hemorrhage, and did post-mortem examinations on 46 eyes of 23 patients with sudden intracranial hypertension.<sup>77</sup> He demonstrated an increased diameter of the optic nerve at the ampullary retrobulbar region compared with controls. Intradural hemorrhages were also commonly seen. In many cases multifocal hemorrhages occurred, and in some the intradural hemorrhage had ruptured into the subdural space.

It has been proposed that optic nerve sheath hemorrhages originate from rupture of intradural vessels and bridging vessels, which run from the dura mater to the pia mater of the optic nerve.<sup>77,95,103</sup> Intraocular hemorrhage results from rupture of the retinal veins secondary to retinal venous hypertension. For this to occur the central retinal vein must be obstructed not only before it communicates with the superior ophthalmic vein, but also at the level of the retinchoroidal anastomosis. In cases of sudden intracranial hypertension, the passage of clear or bloody cerebrospinal fluid (CSF) into the optic nerve sheath subarachnoid space results in compression of the central retinal vein and dilatation of the optic nerve sheath. The retrobulbar region of the optic nerve sheath dilates the most and compresses and obstructs the retinchoroidal anastomosis. For this to occur, the intracranial pressure must rise much higher than the level of pressure rise that results in optic nerve sheath hemorrhage alone.<sup>111</sup>

The rise in pressure in subdural hemorrhages is usually less acute.<sup>80</sup> An acute subdural hematoma can occur in infants with pre-existing chronic subdural hematoma or in subdural effusion.<sup>42</sup> One has been reported in association with Purtscher's retinopathy in an infant with nonaccidental injury.<sup>109</sup> Retinal and preretinal hemor-

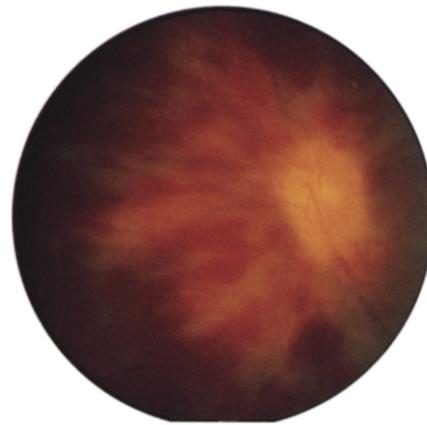


*Fig. 11.* Neonatal retinal hemorrhages occur in up to half of neonates and reabsorb rapidly, usually leaving no sequelae. This neonate shows multiple intraretinal and subhyaloid hemorrhages. (Photograph kindly loaned by Dr. Andrew McCormick, Vancouver, B.C.)

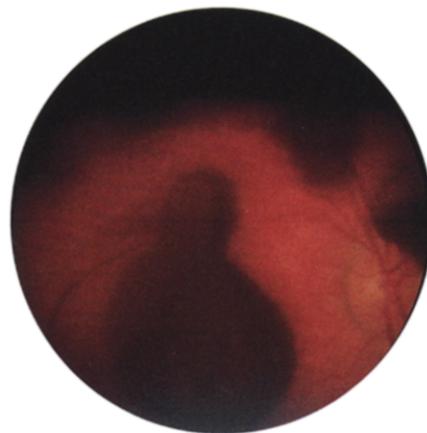
rhages have been noted to be consistently present in infants with acute subdural hematoma.<sup>3</sup> The subdural hematoma is usually parieto-occipital, extending into the interhemispheric fissure posteriorly.<sup>120</sup> Subdural hematoma following accidental head injury is unusual;<sup>38</sup> only one case was found<sup>58</sup> out of 536 children who had accidentally fallen a distance of 20–60 inches.

#### C. RETINAL HEMORRHAGES ASSOCIATED WITH CHILD ABUSE

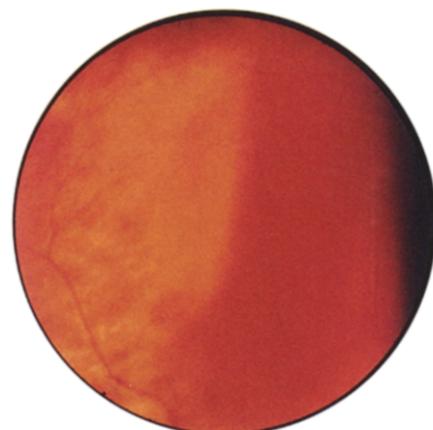
Kiffney (1964)<sup>55</sup> first described the ocular complications in child abuse (nonaccidental injury),



*Fig. 12.* Non-accidental injury (child abuse). There are extensive areas of intraretinal hemorrhage. The optic disc is swollen and pale, the pallor emphasizing the longevity of the disorder. The injury had taken place at least eight weeks previously.



*Fig. 13.* Preretinal hemorrhages in non-accidental injury.



*Fig. 14.* Intraretinal, preretinal and vitreous hemorrhages in non-accidental injury.

of which retinal hemorrhage is the most common. The reported incidence varies from 65% to 89%.<sup>4,11,30</sup> Characteristically, the intraocular hemorrhages are intraretinal, usually involving all the layers of the retina (Fig. 12) and they may last for several months or even years,<sup>48,60</sup> while neonatal retinal hemorrhages usually resolve within a few days.

In more severe cases, in addition to intraretinal hemorrhage, vitreous, subretinal, and preretinal hemorrhages (Fig. 13) have been reported,<sup>81</sup> and mixed forms of hemorrhage are common (Fig. 14). Papilledema occurs secondary to elevated intracranial pressure from subdural hematoma or intracerebral hemorrhage.<sup>118</sup> In child abuse, intraocular hemorrhages are frequently accompanied by intracranial hemorrhages with associated damage.<sup>16</sup> In nonaccidental injury, there are several mechanisms for the causation of retinal hemorrhages.

### 1. Raised Intraocular Venous Pressure

Two mechanisms, often occurring together, are responsible for raised intraocular venous pressure: sudden rise in intracranial pressure and raised central venous pressure.

#### a. Sudden Rise in Intracranial Pressure

Optic nerve sheath hemorrhages may be caused by a sudden increase in intracranial pressure. This could be due to a direct extension of either subarachnoid hemorrhage or cerebrospinal fluid to the subarachnoid space,<sup>60</sup> or it may be secondary to sudden distension of the subarachnoid space from an acute rise in intracranial pressure, which ruptures the dural and bridging

vessels in the optic nerve sheaths.<sup>77</sup> The raised optic nerve sheath pressure compresses the central retinal vein and the chorioretinal anastomosis, thus raising the intraocular venous pressure.<sup>77</sup>

In 1958, Hollenhorst and Stein<sup>41</sup> reported retinal hemorrhages in 51% of infants with intracranial hemorrhages. Muller and Deck<sup>77</sup> noted optic nerve sheath hemorrhages in 87% of eyes examined after a sudden rise in intracranial pressure, but only 27% of these eyes had intraocular hemorrhages. The type and extent of the intraocular hemorrhage depends on the severity of the acute neurologic injury and its resultant rise in intracranial pressure.<sup>118</sup> Smith et al<sup>103</sup> produced preretinal and retinal hemorrhages experimentally in a monkey by acutely raising intracranial pressure with saline infusion into the cisterna magna. A separate subdural hemorrhage of the optic nerve sheath also occurred. They observed retinal venous dilatation immediately when intracranial pressure was increased, indicating acute cerebral and retinal circulatory changes, but they observed the hemorrhage only after a third period of induced pressure rise.

In infants, the vascular system may be particularly susceptible to abrupt changes in pressure. The vessels may be more fragile than in adults, and the normally lower systemic blood pressure, higher cardiac rate, as well as rapid onset of metabolic acidosis with hypoxia, may affect the cerebrovascular hemodynamic forces more readily than in adults.<sup>110</sup> Since cerebral anoxia and ischemia occur secondary to microvascular changes in experimental intracranial hypertension, it is postulated that concomitant retinal an-

oxia and ischemia could occur.<sup>1</sup>

### *b. Raised Central Venous Pressure*

Acute raised central venous pressure can occur in expiration against a closed or partially closed glottis.<sup>60</sup> The Valsalva maneuver, associated with episodes of crying, seizures or squeezing of the chest, or any mechanism where there is a forced contracture of the chest, may cause the so-called Valsalva's hemorrhagic retinopathy.<sup>22</sup>

Purtscher's retinopathy, a hemorrhagic retinal angiopathy, usually results from sudden compression of the thorax or from automobile accidents when there is an acute transmission of high intravascular pressure to the eyes. This can cause angiospasm and subsequent hypoxia of the retina,<sup>68</sup> perhaps associated with arteriolar occlusion from embolization.<sup>14</sup> Purtscher's retinopathy has been reported in nonaccidental injury of children with a history of seizures and chest injury,<sup>109</sup> although it is more commonly recognized in adults. Valsalva's hemorrhagic retinopathy is usually associated with superficial retinal hemorrhages only, while hemorrhages into all layers of the retina may be more common in nonaccidental injury, at least in pathological studies.<sup>60,81</sup> Purtscher's retinopathy is usually accompanied by retinal infarcts and superficial hemorrhages.<sup>68</sup>

The sudden rise in intrathoracic pressure as a result of forceful contraction of the thoraco-abdominal muscles against a closed glottis causes an acute rise in the central venous pressure, which, in turn, increases the intracranial pressure, causing a rise in intraocular retinal venous pressure. This could result from the acute raised central venous pressure occurring with the Valsalva maneuver,<sup>22</sup> traumatic cases, when there is forced compression of the chest usually antero-posteriorly against a closed glottis,<sup>28</sup> epileptic seizures,<sup>2,40,73</sup> vomiting or spells of coughing,<sup>40</sup> especially when they occur simultaneously.

The normal venous pressure values measured in the sagittal sinus in the newborn averaged 6 cm/H<sub>2</sub>O;<sup>85,113</sup> it varied instantaneously up to 20 cm/H<sub>2</sub>O during crying, abdominal straining, and convulsions. Intubated patients who are obviously incapable of Valsalva's maneuver showed a minimal and variable rise in venous pressure of 2–3 cm/H<sub>2</sub>O during crying or convulsions.<sup>113</sup> During generalized tonic seizures in status epilepticus, the pressure reached 18–23 cm/H<sub>2</sub>O.<sup>113</sup> In experimental animals, values of cerebral venous pressure above 100 cm/H<sub>2</sub>O during a tonic seizure have been demonstrated.<sup>113</sup> The increase

in muscle tone and intrathoracic pressure makes an important contribution to this;<sup>72</sup> thus, transient rises accompany whole body jerks during a myoclonic seizure. Cerebral vasodilatation is also an important factor, as seen in paralyzed, artificially ventilated animals.<sup>73</sup> These marked transient rises in cerebral venous pressure in association with simultaneous changes in arterial blood pressure result in pronounced alterations in perfusion pressure.<sup>85</sup> These impair autoregulation<sup>64</sup> following a rise in intrathoracic pressure and are probably responsible for the small cerebral hemorrhages seen after experimental seizures.<sup>39,59,74,85</sup> Retinal hemorrhages may occur under these circumstances, but only rarely do they have more than transient effect.<sup>40,51</sup> The coincident rise in intraocular pressure at such times reduces the pressure difference across the vessel wall and may give a protective effect against more profuse bleeding.<sup>40</sup>

A similar event may occur transiently when an open airway is exposed to very high pressure. One situation in which this may occur is in resuscitation. Normally in resuscitation after a cardio-pulmonary arrest, under the relatively controlled circumstances in hospital, there is a good airway and the relatively skilled resuscitator uses enough exhalation pressure to inflate the lungs and there is no resistance by the patient. Retinal hemorrhages were not present under such circumstances except in one patient who had seizures and arterial hypertension after cardiopulmonary resuscitation without a preceding history of trauma or child abuse.<sup>51</sup>

On the other hand, an inexperienced person – or even more so, a completely nonexperienced and panicking person – would be likely to overinflate the lungs. If a child were unconscious and not able to resist the exhaled air pressure, his chest would expand even to the extent of causing some lung damage, but if he is semiconscious and struggling, his chest wall and diaphragm will be rigid and the exhaled air from the resuscitator will enter his lungs at a very high pressure. This, combined with general muscular contracture, would probably cause pulses of very high central venous pressure.

## **2. Rapid Deceleration or Cycles of Rapid Acceleration and Deceleration**

Lyle et al (1957)<sup>65</sup> have shown that rapid deceleration alone can produce retinal hemorrhages. The incidence for this was found in human volunteers subjected to high deceleration forces (negative acceleration). Experiments using rock-

et-propelled sleds that were decelerated by a braking system reproduced the forces encountered in supersonic escape from aircraft.<sup>65</sup> Retinal hemorrhages were produced in subjects exposed to forces as great as 40G for one second, and the fundus lesions were thought to be caused by combined hydrostatic and decelerative forces. Similar fundus changes have been observed in pilots following bailout from supersonic aircraft.

In 1980, Ober<sup>81</sup> proposed that the rapid acceleration and deceleration associated with the shaking of infants' heads accounted for the hemorrhagic retinopathy, but Duhami et al (1987)<sup>23</sup> suggested that severe examples of the shaken baby syndrome are not usually caused by these forces alone. The tendency for a more severe retinopathy in infants could be explained by clinical and experimental evidence, which suggests that intraocular hemorrhages may occur from abrupt elevation in retinal venous pressure secondary to an acute rapid rise in intracranial pressure.<sup>60,77</sup>

Caffey<sup>15</sup> noted the association between retinal hemorrhages and intracranial injuries in shaken infants, and suggested that the head of an infant is particularly vulnerable to whiplash injuries due to the proportionately larger and unsupported head, the pliability of the sutures and fontanelles that allows stretching of the calvarium, the greater deformability of the unmyelinated brain, and the greater percentage of cerebrospinal fluid. All these features of an infant's head allow greater shearing stresses to be exerted on the intracranial blood vessels. Shaking may, therefore, cause generalized brain contusion. Alternatively, bilateral subdural hematomas may be induced and a posterior interhemispheric distribution is particularly characteristic of shaking injury. These factors may contribute to the more severe acute elevations of retinal venous pressure, which result in bilateral retinal hemorrhages in the absence of external signs of head injury.<sup>15</sup> Zimmerman et al<sup>120</sup> found 65% of 26 children with nonaccidental injury had retinal hemorrhages; this represented 80% of those with interhemispheric subdural hemorrhage. The presence of retinal hemorrhages with localized intracerebral hematoma is most unlikely to be traumatic if there is no evidence of parenchymal injury.<sup>72</sup>

The retinal hemorrhages usually extend throughout all layers of the retina<sup>78</sup> and into the subretinal space, and may result in permanent visual handicaps due to macular scarring,<sup>37</sup> retinal detachment,<sup>116</sup> optic atrophy,<sup>36</sup> and retino-

schisis.<sup>35</sup> In the shaken baby syndrome, Greenwald and associates<sup>35</sup> reported a series of infants with retinal cysts filled with blood or clear fluid as in retinoschisis. The vision was poor when the macula was involved. They suggested that head-shaking placed traction on the retina by transmission through the lens and vitreous humor, causing hemorrhage and cystic lesions. Sometimes, attempted resuscitation of a lethargic child by shaking the infant causes to and fro flexions of the head and may result in intracranial and intraocular hemorrhages and death.<sup>1</sup>

Perimacular folds may be characteristic of shaken babies without direct head trauma; they are white, ring-shaped folds which encircle the macula outside the vascular arcade.<sup>32</sup> The folds are probably caused by vitreous traction; the vitreous is attached at the apices of the folds and detached in their bases.<sup>70</sup> The perimacular folds may be suggestive of shaking alone, without direct head trauma, and this is always a nonaccidental injury.<sup>70</sup> The presence of direct head trauma, which may also occur in accidental injury, however, may be subtle and detectable only at autopsy.<sup>25</sup>

## D. RETINAL DISEASES

### 1. Coats' Disease

Coats' disease is a rare form of retinal telangiectasia with secondary subretinal exudation that may present in infancy. It is frequently unilateral. The walls of these telangiectatic retinal vessels are abnormal with resultant transudation of fluid and retinal hemorrhages.<sup>87</sup>

### 2. Persistent Hyperplastic Primary Vitreous (PHPV)

PHPV is a developmental anomaly that may present with microphthalmos, retrothalamic mass, vitreous hemorrhage, and secondary glaucoma which is very difficult to treat.<sup>52</sup> Intralental hemorrhages have also been described.

### 3. Retinopathy of Prematurity

The occurrence of retinal hemorrhage in the acute phases of the disease process has been recognized for many years. Usually these retinal hemorrhages are localized on the surface of the neovascular ridge at the junction of vascularized or nonvascularized premature retina. Frequently, the hemorrhages are small and restricted to the demarcation zone. Occasionally, they may be massive and extend into the vitreous. Vitreous hemorrhage may be associated with progression

of disease.<sup>97</sup>

Retinal hemorrhages have been found to be more frequent in infants treated with parenteral vitamin E (tocopherol). In a double-masked, randomized, placebo-controlled trial of early parenteral tocopherol given from birth to 287 infants, retinal hemorrhages correlated strongly with plasma tocopherol levels from three weeks to three months. Coagulation studies were not done routinely in these patients.

Ehrlich et al<sup>24</sup> demonstrated that tocopherol treatment impairs the deposition of collagen in healing wounds in rats, and delays growth of new vessels into the subcutaneously implanted sponges. Such effects in the eye could allow poorly supported vessels to bleed more readily.

#### **4. Retinal Dysplasia**

Retinal dysplasia is most commonly seen in patients with Norrie's disease. This X-linked recessive syndrome presents in the first few months of life with retinal dysplasia associated with extensive hemorrhage.

#### **5. Hypertension**

In extremely premature infants, intracerebral hemorrhage is a major complication and there has been concern that hypertension may be a factor. Transient increases in blood pressure during crying may be a problem.<sup>87</sup> Hypertensive retinopathy in infants with associated renal problems may, rarely, be acute and malignant.

#### **6. Myopia**

Pathologic myopia usually has its onset at birth or in the neonatal growth period. The earlier the onset of myopia, the greater will be the severity and progression.<sup>12</sup> Infants with high myopia are, therefore, at risk of developing retinal hemorrhages secondary to chorioretinal changes such as lacquer cracks and choroidal neovascularization.

#### **E. BLOOD DYSCRASIAS**

Retinal hemorrhages frequently occur in blood dyscrasias with pancytopenia, anemia, and coagulopathies, such as those associated with acute lymphoblastic leukemia. They have also been described in megaloblastic anemia with vitamin C, vitamin E and vitamin K deficiency states.<sup>8,49,87</sup>

In leukemia, retinal hemorrhages occur as a result of a combination of infiltration and damage to retinal vessel wall, poor coagulation, hyperviscosity, and vessel occlusion. Hemorrhages

occur throughout the retina and usually involve all the layers of the retina and may also involve the vitreous. In addition, white-centered hemorrhages (Roth's spots), "cotton wool" spots, and retinal infarcts occur frequently in acute leukemia.<sup>56,121</sup> In white-centered hemorrhages, the white area consists of platelet and fibrin deposits or septic emboli.<sup>21</sup>

In sickle cell disease, pre- or intraretinal hemorrhages occur due to extravasation of blood through the damaged arteriolar wall following transient arteriolar occlusions and subsequent reperfusions. Talbot et al<sup>105</sup> saw these changes in the youngest children that they examined in their cohort (five years old). In the opinion of these authors, retinal hemorrhages could occur from the time when HbS becomes predominant in the infant's blood, and they believe that retinal hemorrhages could occur as young as one year of age in some affected infants (Talbot JF: Personal communication).

#### **F. BLOOD HYPERVISCOSITY**

Retinal hemorrhages occur in patients with blood hyperviscosity, such as in cystic fibrosis,<sup>91</sup> macroglobulinemia,<sup>82</sup> cryoglobulinemia,<sup>18</sup> and paraproteinemia.<sup>18</sup> The mechanism by which these scattered retinal hemorrhages develop is not clear. In cystic fibrosis, hemorrhages have been found in patients with moderate to severe pulmonary disease<sup>91</sup>; in this instance the mechanism probably includes a combination of hyperviscosity, hypoxia and hypercapnia.

#### **G. HYPOTENSION AND ANOXIA**

Retinal and optic nerve ischemia and hemorrhages may be caused by sudden reduction of systemic blood pressure because of cardiac decompensation, traumatic shock, or therapy for arterial hypertension.<sup>84</sup> Sudden surgical lowering of intraocular pressure (e.g., glaucoma filtering surgery) in the presence of a healthy retinal vessel seldom produces retinal bleeding, but anoxia together with increased transmural pressure is very likely to produce bleeding, e.g., following difficult breech deliveries or deliveries during which the cord is strangulating the neck.<sup>78</sup>

#### **H. INFECTIONS**

Retinal hemorrhages can occur in viral infections, such as cytomegalovirus (CMV) and herpes simplex, rickettsial infections,<sup>34</sup> ocular toxoplasmosis, and subacute bacterial endocarditis.

### **1. Cytomegalovirus (CMV)**

CMV retinopathy is characterized by full-thickness necrosis of the retina, which is seen as discrete white patches with feathered and indistinct borders similar to cotton-wool spots and flame-shaped hemorrhages resembling crumbled cheese and ketchup. Retinal involvement usually follows the vascular distribution, producing large areas of necrosis or smaller perivascular infiltrates. The lesions of CMV retinitis progress slowly, taking perhaps a month to double or triple in size.<sup>100</sup> It usually begins in the posterior pole, although it may involve any part of the retina. CMV retinitis can occur in children with AIDS<sup>100</sup> or neoplastic disease,<sup>9</sup> and in children who have received cytotoxic or corticosteroid therapy.

### **2. Herpes Simplex**

Retinopathy is the most serious of the disorders associated with herpes simplex virus, which causes retinal necrosis seen as patches of retinal whitening on ophthalmoscopic examination, and vasculitis.<sup>87</sup> Retinitis may not develop until many months after birth.<sup>31</sup> The virus reaches the retina by neural spread from the brain or by hematogenous spread.<sup>87</sup>

### **I. CAROTID-CAVERNOUS FISTULA**

Carotid-cavernous fistula, usually traumatic in origin, is vastly less common in infancy than in adults.<sup>17</sup> The fistula produces hemodynamic changes resulting in a lowered ophthalmic arterial pressure and a raised venous pressure in the eye and orbit. Prolonged reduction in perfusion pressure and ischemia may result in widespread intraretinal and superficial hemorrhages.<sup>96</sup>

### **J. FAMILIAL RETINAL ARTERIOLAR TORTUOSITY**

This progressive autosomal dominantly-inherited syndrome of retinal arteriolar tortuosity may be complicated by spontaneous retinal hemorrhages,<sup>117</sup> which usually resolve spontaneously. It affects small retinal arterioles mainly in the macular area, which may be overlooked.

### **K. PROTEIN C DEFICIENCY**

Protein C deficiency is an autosomal recessive disorder. Heterozygous individuals may have an increased disposition to recurrent venous thrombosis, whereas homozygous individuals may develop widespread thrombotic complications during the neonatal period. Subarachnoid hemorrhage with intravitreal hemorrhages have

been reported in a boy born at 37 weeks gestation by normal delivery.<sup>86</sup> The protein C level of the neonate was less than 5% (normal 99.6%) and both parents had low levels also (40%). Vitreous hemorrhage in a neonate, when associated with other signs of reduced clotting ability, should alert the clinician to test for protein C activity.

### **L. OTHER CAUSES**

Rarely, vitreous hemorrhage may be seen at the onset of retinoblastoma, and, in fact, it may be the presenting sign. Vitreous hemorrhage may also occur with optic disc hamartoma (mulberry tumors), tuberous sclerosis, and juvenile X-linked retinoschisis. Other predisposing factors include drugs (e.g., aspirin, anticoagulants), hypercapnia, coagulation defects, and trauma.

## **IV. Summary**

Intraocular hemorrhages can be classified according to their anatomic location and morphological appearances, e.g., intravitreal, preretinal, intraretinal, subretinal, subretinal pigment epithelial, choroidal, and white-centered.

Neonatal hemorrhages are very common, intraretinal, and they resorb rapidly and usually completely. Fundus hemorrhages commonly occur with subdural and subarachnoid intracranial hemorrhages. They are peripapillary, sometimes extensive, breaking into the vitreous, and reabsorption may be delayed.

Retinal hemorrhages occur in up to 89% of infants who have suffered child abuse (nonaccidental injury), and the more severe the hemorrhages, the more extensive the intracranial pathology. Perimacular folds may be pathognomonic of child abuse by shaking, but retinal hemorrhages cannot by themselves be taken as being pathognomonic of child abuse. Retinal hemorrhages in child abuse may affect all layers of the retina, but they are frequently preretinal and breaking into the vitreous.

Pathogenetic mechanisms include raised intraocular venous pressure secondary to raised intracranial pressure and raised central venous pressure, as well as rapid deceleration or cycles of rapid acceleration-deceleration.

Retinal hemorrhages are frequently seen in retinopathy of prematurity, especially with advancing disease. In leukemia and other blood disorders, retinal vascular disease with hemorrhage is very frequent. Rare causes of retinal hemorrhage in infancy include Coats' disease, PHPV, retinal dysplasia, hypertension, hypervis-

cosity, retinal infections, hypotension and anoxia, carotid-cavernous fistula, Protein C deficiency, high myopia, and a rare dominantly inherited syndrome of retinal arteriolar tortuosity with spontaneous retinal hemorrhages.

Early detection and management of retinal hemorrhages in infancy, especially macular and vitreous hemorrhages, may be essential to prevent irreversible visual loss.

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## Outline

- I. Clinical appearance
- II. Pathological anatomy
  - A. Vitreous hemorrhages
  - B. Preretinal hemorrhages
  - C. Intraretinal hemorrhages
  - D. Subretinal hemorrhages
  - E. Subretinal pigment epithelial hemorrhages
  - F. Choroidal hemorrhages
  - G. White centered hemorrhages
- III. Associated conditions and etiology
  - A. Neonatal hemorrhages
  - B. Subarachnoid and subdural hemorrhages
  - C. Retinal hemorrhages associated with child abuse
    1. Raised intraocular venous pressure
      - a. Sudden rise in intracranial pressure
      - b. Raised central venous pressure
    2. Rapid deceleration or cycles of rapid acceleration and deceleration
  - D. Retinal diseases
    1. Coats' disease
    2. Persistent hyperplastic primary vitreous
    3. Retinopathy of prematurity
    4. Retinal dysplasia
    5. Hypertension
    6. Myopia
  - E. Blood dyscrasias
  - F. Blood hyperviscosity
  - G. Hypotension and anoxia
  - H. Infections
    1. Cytomegalovirus
    2. Herpes simplex
  - I. Carotid-cavernous fistula
  - J. Familial retinal arteriolar tortuosity
  - K. Protein C deficiency
  - L. Other causes
- IV. Summary

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