The Treatment of Hemangiomas:

With Special Reference to the Role of Steroid Therapy

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Hemangiomas are the most common of all human birth defects. The author has reviewed a 25-year personal experience with treatment of over 1000 patients with a variety of common and rare developmental vascular anomalies. Attention is given to a more useful clinical classification of these disorders. The classification is intended to be helpful in estimating prognosis of the lesions and as a guide to the choice of therapy. Many treatment modalities are evaluated—some successful and some quite disappointing. Surgical excision, irradiation, CO_2 freezing, sclerosing agents, cauterization, steroid therapy and watchful waiting are among the treatment methods evaluated. High dose—short course Prednisone therapy has proved to be a major new addition to the treatment of massive juvenile capillary hemangiomas. Numerous misconceptions have appeared in the medical literature. These are noted and a philosophic basis for present day management is suggested for each type angioma. The roles of growth, resolution, histologic picture and sense of deformity are considered in viewing the surgeon's approach to these difficult and challenging problems. Some evidence and speculations are offered as to the etiology, neurogenic influences and physiological dynamics of the various hemangiomas.

HEMANGIOMAS are the most common congenital anomalies in humans and they present physicians with some of the most difficult treatment problems known to medicine.

Hemangiomas are *not* true neoplasms despite the fact that they have the potential to grow. They might be defined as congenital hamartomas, representing mesodermal rests of vaso-formative tissue. As they enlarge, neo-vascularization occurs by canalization of solid masses of endothelial cells.

There is much confusion and disagreement in the medical literature concerning the nature and management of hemangiomas. This confusion stems from the fact that the term "hemangioma" has been used loosely in medical literature for many years to include a great variety of very diverse clinical conditions. Classification of angiomas is further complicated by the marked limitations in the ability of the medical pathologist to separate the clinical varieties of hemangiomas on the basis of histologic sections. All hemangiomas have abnormal patterns of hemodynamics, and these *physiological* differences, not seen in fixed sections, are largely responsible for the great clinical variations in different varieties of hemangiomas.

The prognosis of two different types of hemangioma may vary from rapid and complete involution to continous progression leading to ulceration, infection, heart failure and even death. The choice of the optimum method of treatment for a given hemangioma not only depends on the specific variety of hemangioma under consideration, but also may change with its precise extent, the age of the patient and the exact anatomical location of the angioma. Both treatment and prognosis of hemangiomas are further complicated by the fact that the congenitally predetermined ultimate extent and pattern of a given hemangioma may be occult and completely indeterminate at the time of initial diagnosis. A workable classification of hemangiomas should provide guidance as to the most appropriate treatment program to be followed (Table 1).

Over the past 25 years the author has personally dealt with several thousand hemangiomas, including examples of all categories listed in Table 1. A large variety of proposed treatment methods have been tried

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TABLE 1. *Classification of Hemangiomas*

Type 1: Neonatal Staining
Type 2: Intradermal Capillary Hemangiomas
A. Salmon Patch
B. Port Wine Stain
C. Spider Angiomas
Type 3: Juvenile Hemangiomas
A. Strawberry Mark
B. Strawberry Capillary Hemangioma
C. Capillary Cavernous Hemangioma
Type 4: Arteriovenous Fistulae
A. Arterial Hemangiomas
B. Hemangiomatous Giantism
Type 5: Cirsoid Angioma (racemose aneurysm)

when appropriate indications existed. A number of lessons have been learned concerning the benefits and limitations of these treatment methods. This paper will attempt to summarize (and possibly oversimplify) the basic principles of selection and timing of treatment approaches for each of the principle varieties of hemangioma.

Treatment of Hemangiomas Based on Clinical Classification

Type 1: Neonatal Staining

This condition is present at birth and is seen as a light pink staining pattern of the skin usually located in the midline of the base of the neck over the ligamentum nuchae, or over the glabella region of the forehead. It is occasionally seen over the sacrum. This condition almost always fades spontaneously with a few months, and no treatment is indicated.

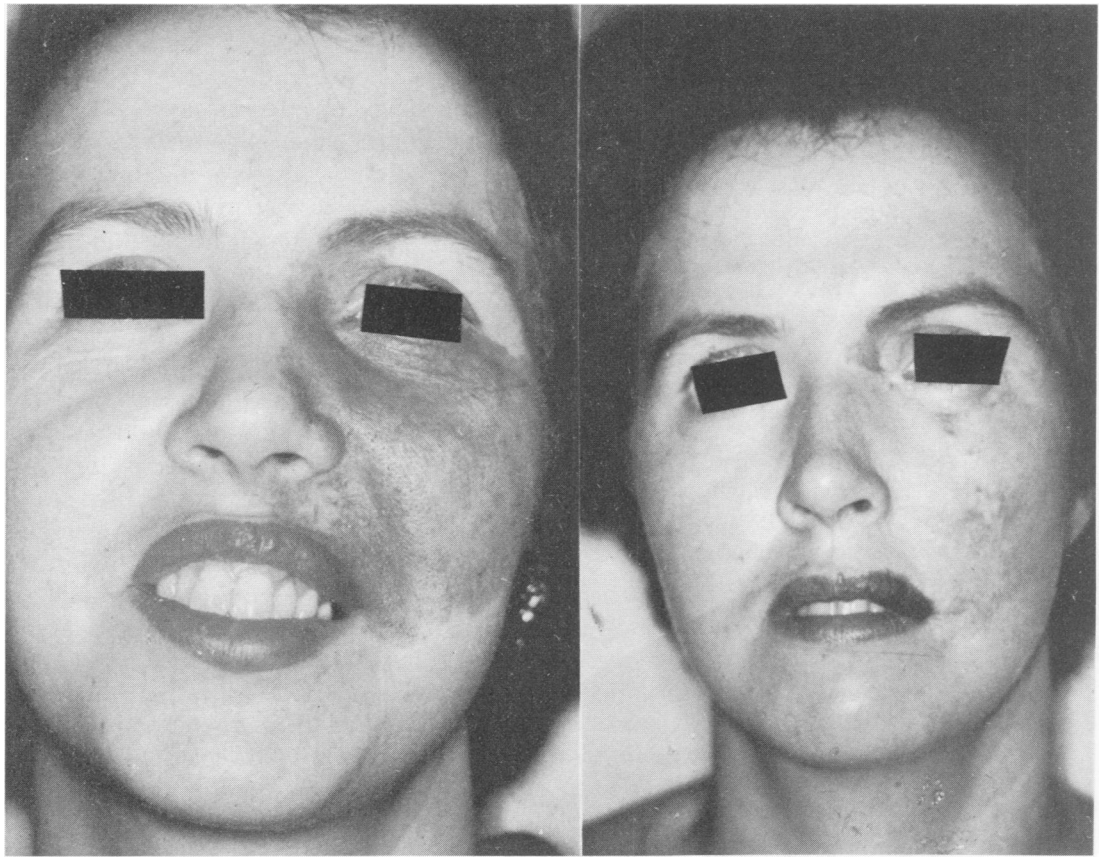
Type 2: Intradermal Capillary Hemangiomas

These are usually present at birth. The endothelium shows adult type morphology and is located in the deep dermis. The discoloration patterns correspond to the skin distribution of various branches of sensory nerves. On the face, they most commonly reflect patterns of the trigeminal nerve branches. There are three varieties:

The salmon patch: This is an extremely faint variety of intradermal hemangioma with a light pink to rust color. It blanches on pressure, is not elevated above the skin surface, and its pattern does not change over a period of years. It is not sensitive to x-ray therapy, surface dessication, freezing, surgical shaving, or abrasion.

Treatment consists of no treatment, use of make-up creams (in females only), or medical tattooing with insoluble pigments. In very exceptional cases, removal of the salmon patch and skin grafting may be indicated when the sense of deformity is marked.

Unfortunately, tattooing requires multiple treatments, often over a period of years, and although the color may



FIGS. 1a and b. (left) Young school teacher with port wine stain involving skin innervated by maxillary division of left trigeminal nerve. She had tried to cosmetically cover the area for several years but found this tedious and stated "it did not make me feel any less disfigured." Radioactive thorium had been tried but proved unsuccesful. (right) One-stage massive skin grafting of the left cheek, left nose, left lower eyelid, and left upper lip was carried out in a single operation. The skin graft was taken from the anterior and lateral aspect of the left neck at a setting of 0.012 of an inch. Only the light port wine stain in the left upper medial eyelid and that within the vermillion of the lip were left. Patient no longer uses make-up of any kind and has found the removal of the birthmark "most gratifying."

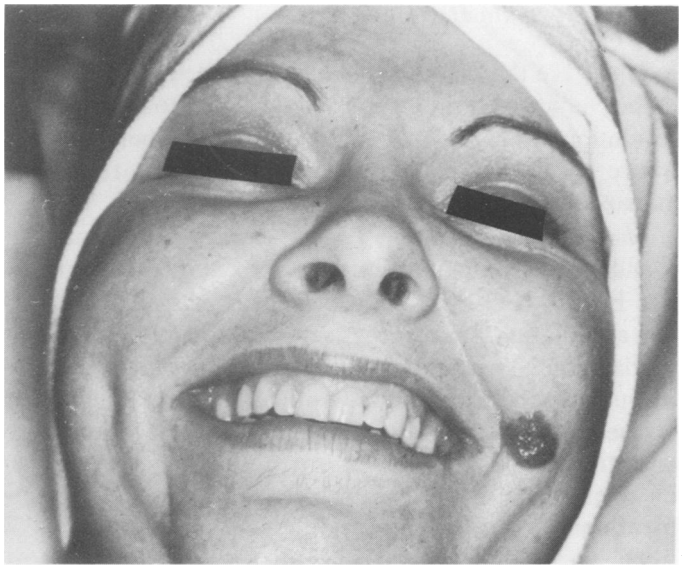


FIG. 2a. This young nurse's mother had been told 20 years earlier that this capillary cavernous hemangioma would "disappear if she would give it time." At 20 years of age, the patient is still waiting.

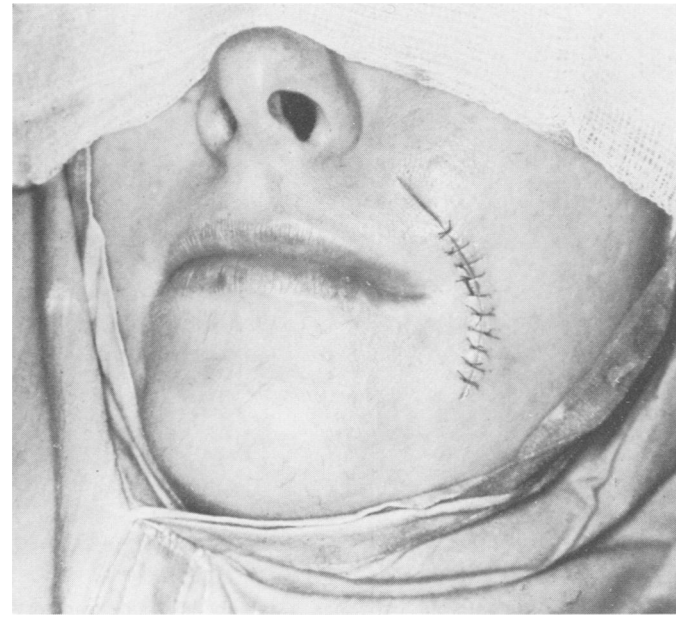


FIG. 2b. This localized hemangioma is removed using a natural skin line of relaxation with plastic closure.

be improved it has not been possible to provide skin tones that will match the changes in the color of the normal surrounding skin. Tattooed angiomas do not show appropriate color variations with changes in temperature and external light reflection. A tan or brownish patch-like effect is often the result.

Port wine stain. The port wine stain is much deeper in color than the salmon patch. It does not enlarge but, with time, often develops hyperkeratotic patches or knobs on its surface. These are similar to changes seen in certain types of pigmented nevi and probably represent abnormalities of cutaneous nerve endings. After many years, the surface of port wine stains may develop a troublesome eczema if untreated.

Treatment: When the port wine stain involves a major area of the face, skin grafting is probably indicated (Figs. 1A and B). Grafts should be patterned to match esthetic facial units to make marginal scarring less conspicuous. Skin grafts should be taken from color-matched donor sites (the scalp or posterior neck) that normally have capillary beds capable of blush responses. This type of grafting must be executed with flawless technique for good results. It is more difficult to obtain perfect results in very young children, but grafting should not be postponed until adult life as *much psychologic damage occurs during the waiting period!* The sense of deformity with a port wine stain is much more consistently present and profound than is generally appreciated.

Spider angiomas. These have a small central dermal arteriole with a network of radiating intradermal capillaries reachiang out in stellate fashion. They are small—but may be multiple—and they often appear at 3 to 4 years

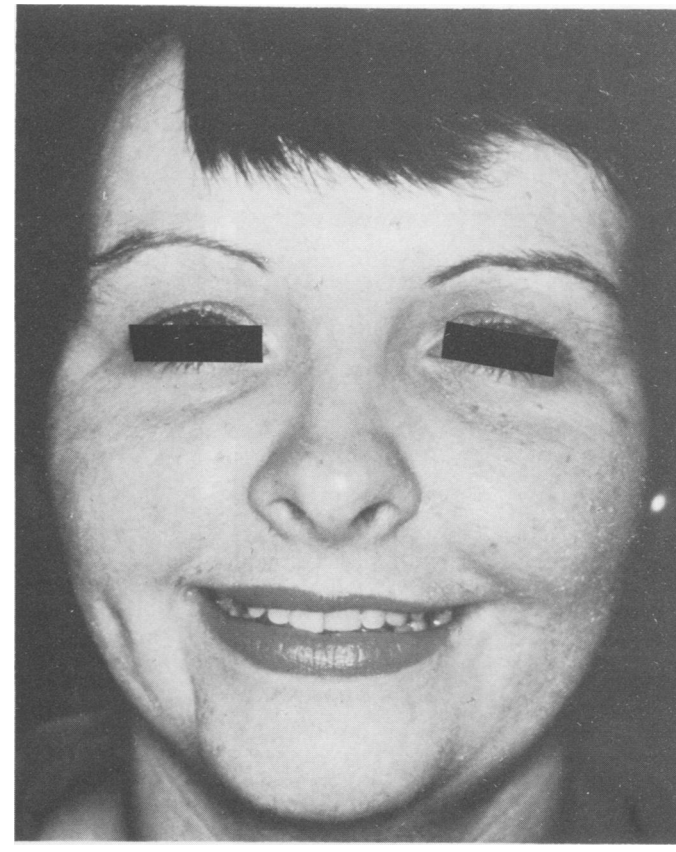


FIG. 2c. This shows the result obtained some months later, the procedure having been carried out under local anesthesia as an outpatient procedure. If the same treatment had been used in childhood, many years of embarrassment would have been avoided.

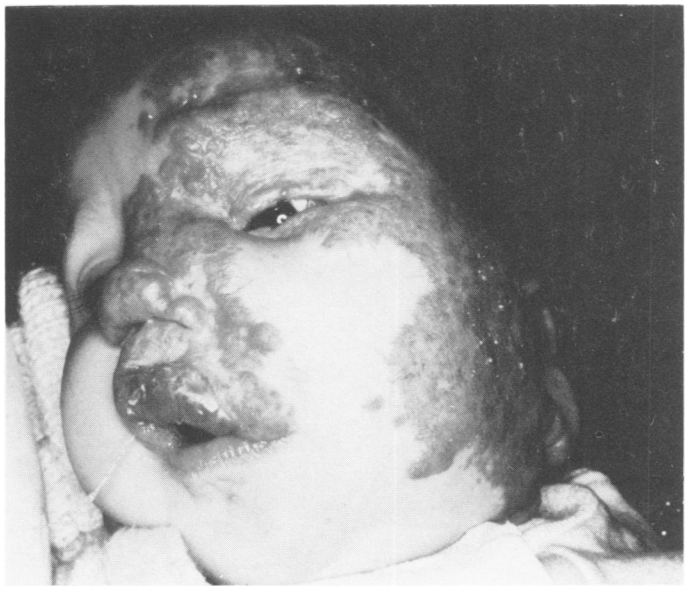


FIG. 3a. This 2-month-old baby has a rapidly enlarging capillary cavernous juvenile hemangioma. The pediatrician seeing this baby urged "watchful waiting."

of age. The pulsations of the central arteriole may be brought out by pressing a glass plate lightly over the spider. They usually require several years for spontaneous resolution.

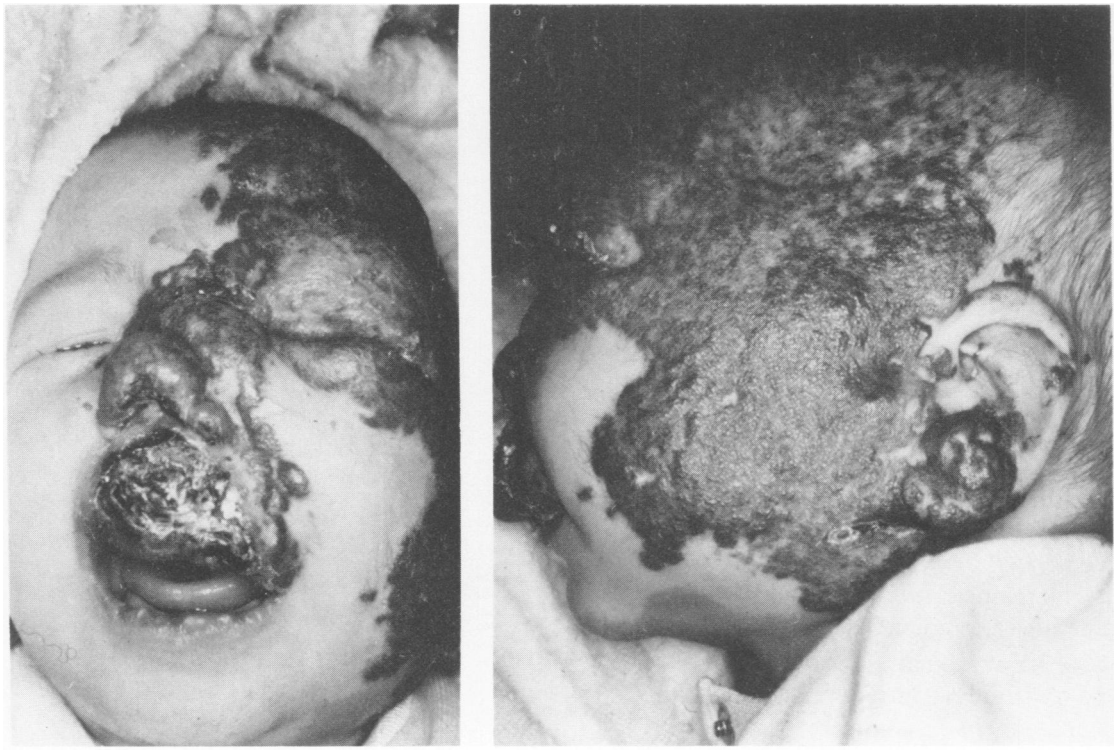
Treatment: If resolution is unduly delayed, they may be treated by fine needle point dessication of just the central arteriole, using magnification.

Type 3: Juvenile Hemangiomas

These are the common lesions described so frequently in the pediatric literature as "always undergoing complete resolution." They represent the only type of hemangioma where resolution plays a significant role in treatment management. There are three varieties:

The strawberry mark. The strawberry mark may be found in approximately one out of every one hundred live births. It consists of a pale halo of skin surrounding a radiating telangiectasis approximately 1–3.5 cm in diameter. The halo may be brought out more clearly by rubbing the skin surface with the finger. Since most of these lesions will disappear spontaneously, they are sometimes thought of as "abortive" angiomas. A few go on to produce a "strawberry mark *en plaque*." Treatment consists of watchful waiting and reassurance.

The strawberry capillary hemangioma. This is perhaps the most common congenital anomaly in children. It may be present at birth but often appears as a tiny red spot in the first days or weeks of life and will grow rapidly in size over the first three months of life. When limited to the surface capillaries of the skin, it may be relatively circumscribed, but is usually moderately elevated above the surface with some degree of lobulation and a bright red color that gives it the name "strawberry." These smaller lesions rarely ulcerate and in the vast majority of instances will undergo spontaneous resolution between about one and 4 years of age. After age 4,



FIGS. 3b and 3c. Within 2 weeks the lesion enlarged to produce ulceration of the upper lip, loss of the columella of the nose, closure of the left eye, blockage of the left ear canal with otitis externa and abscess and drainage from the left parotid gland. The patient developed fever, anemia, and stridor.

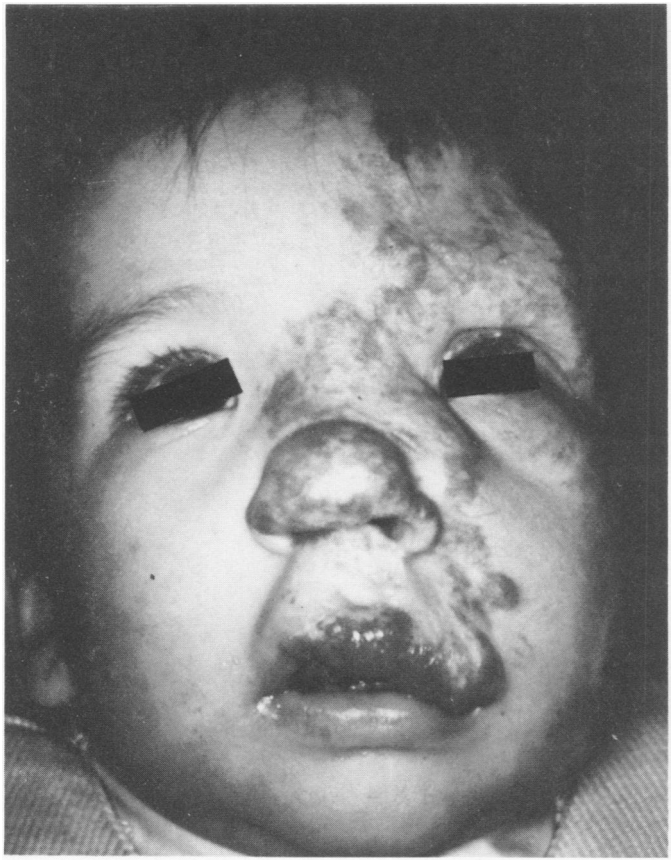


FIG. 3d. Patient was placed on 40 mg of Prednisone q/2 days when seen in the condition of Figs. 3B and 3C. Within 2 weeks there was marked reduction in the size of the lesion and the ulcerations had begun to heal.

spontaneous regression may continue—but only at an extremely slow rate.

Treatment: When these lesions are relatively small and on noncritical areas of the body, spontaneous resolution may give satisfactory results. If ulceration occurs because of irritation or involvement of the surface epithelium, infection and permanent scarring are inevitable. With such lesions early surgical excision may offer the best solution for both child and parents. Even non-ulcerated strawberry hemangiomas may, in many instances, be most effectively managed by simple excision and skin closure (Figs. 2a to c). Modern techniques of plastic skin repair make it possible to greatly minimize surgical scarring and avoid years of deformity and concern about these lesions. To await complete resolution is a disservice to both child and parents in the case of lesions well suited for surgical excision. Irradiation therapy or freezing techniques that will produce ulceration and scarring are contraindicated. Sclerosing agents such as 5% sodium morrhuate (a mixture of the sodium salts in the saturated and unsaturated fatty acids of cod liver oil) should be limited to injections of lesions on the scalp, tongue, buccal surface and other areas where residual scarring is of little concern and surgery may be unnecessary. In these special areas a mechanical device may be used to arrest circulation in the injected area for about 5 minutes to greatly improve the response and sclerosis.

Capillary cavernous hemangiomas: At times juvenile capillary hemangiomas involve not only the cutaneous vessels but also larger venous sinusoids in the deep dermis and subcutaneous tissues. Such hemangiomas may take on gigantic proportions involving major portions of an extremity or several features of a child's face and neck. The growth pattern (most rapid in first 6 months of life) is very similar to that of its smaller cousin, the strawberry capillary hemangioma; but because of its large size and rapid spread, the lesion may progressively involve normal and vital structures with alarming rapidity. Over a period of a few weeks, eyelids, lip vermillion, ear canals, nasal cartilages, and other facial features may be seen to disappear into the elevated and spreading bright red margins of this tumor. Maceration, ulceration, infection, bleeding, scarring, and functional problems related to breathing, vision, hearing and eating may rapidly ensue. If the infection is controlled by antibiotics and the anemia is corrected, spontaneous resolution will occur in such patients, but it is *never complete* and usually is delayed in age of onset in comparison with the smaller more circumscribed juvenile strawberry angiomas. When parents are advised to have no treatment for this lesion and to wait for resolution, they may be dismayed as the days pass while they see enlarging ulcerations with serious scarring, sepsis, fever, anemia and other com-

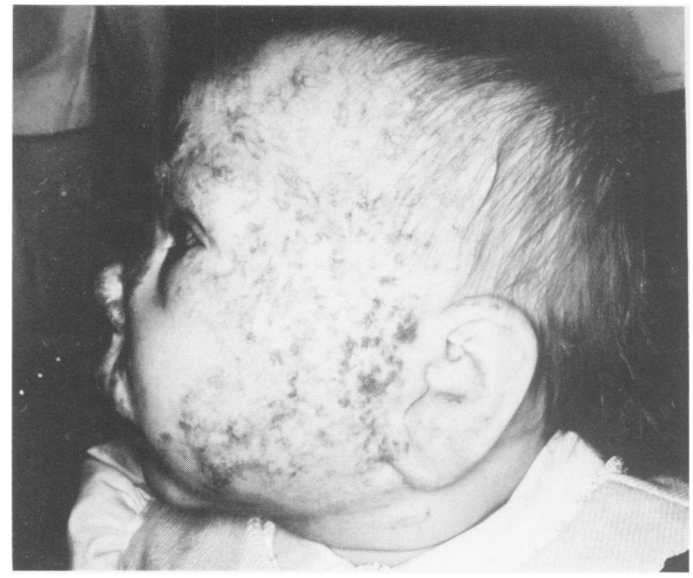
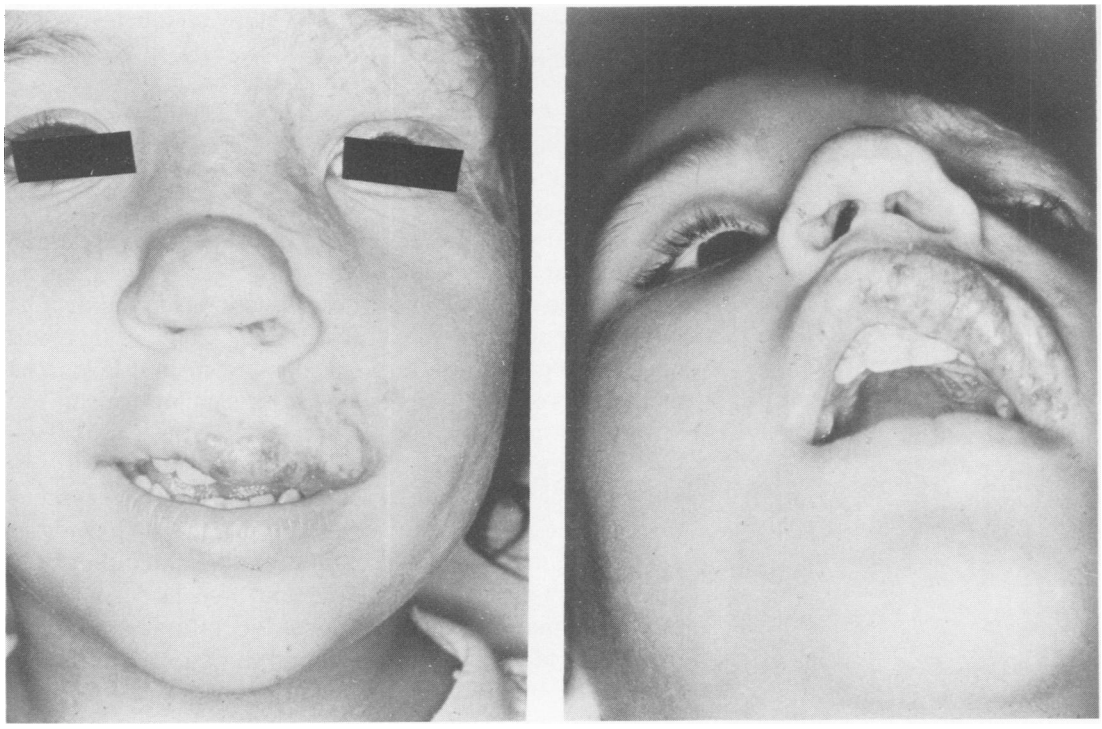


FIG. 3e. Within 5 weeks the condition had markedly improved. The eyes are opened, the ulcerations healed and the deep mass is subsiding in volume. The ear canal is opened and vasospasm and sclerosis may be seen on the left half of the face.



FIGS. 3f and g. At age 5 further resolution of this hemangiom (shown in Figs. 3a through c) had come to a halt for over one year. She was left with residual deep angioma in the upper lip, nasal tip, and left upper eyelid, and with considerable scarring as well as loss of the columella of the nose. Reconstructive surgery was indicated.

plications (Figs. 3a to 3d). Years later children who have been managed by such conservative routines are left with massive deformity. It may be equally unwise and deforming to undertake major surgical resection of these hemangiomas *during the phase of rapid enlargement!* Better treatment of these lesions is now possible.

Role of Steroids in Treatment of Capillary-cavernous Hemangiomas

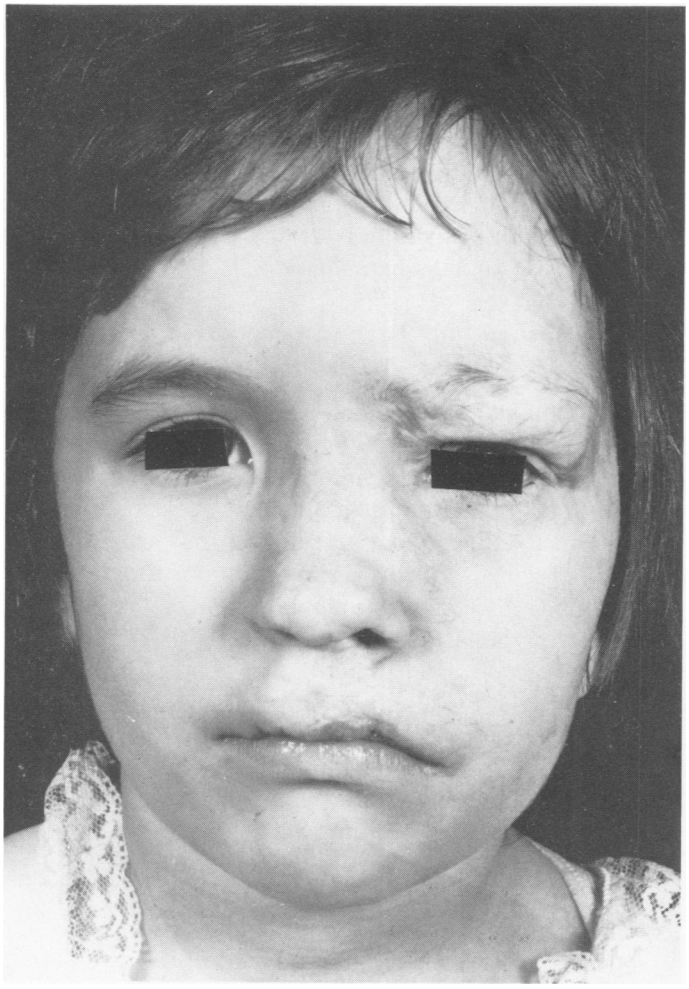
Treatment of the rapidly growing capillary cavernous hemangiomas requires the initiation of a course of steroids (Prednisone) at the earliest possible age. In 1967 we reported the first 7 children who showed uniform and dramatic responses of their massive capillary cavernous hemangiomas to "short course—heavy dose" steroid therapy.[23] Earlier attempts had been made to manage the thrombocytopenia of infants with Kassabach-Merritt Syndrome by the use of steroids, but results had been somewhat disappointing. In a few of those children, the size of the hemangioma had decreased, but platelet levels had not been corrected. Subsequently, Fost and Esterly[6] and Brown, *et al.*[3] have reported confirmation of the gratifying results seen[23] in infants with massive juvenile capillary hemangiomas following treatment with Prednisone.

Since 1967 the author (M.T.E.) has continued to use steroids with this group of patients with a better than 90% dramatic response of these lesions as manifested by marked regression within two to three weeks after starting therapy. With daily (or 1. 2 day) doses of 20–40 mg of Prednisone, one may expect the enlarging hemangioma to stop growing in 3 to 21 days. Ulcerations already present will heal within two weeks. Shrinkage of the lesion will usually follow if the treatment is continued for 30–90 days. If there is laryngeal involvement and stridor, this is usually dramatically relieved. If, on stopping the steroids, there should be any rebound of growth, a second or third course of hormone will usually be effective.

Alternate day steroid therapy may reduce the likelihood of steroid dependency and minimize complications. However, this remains unproved and several infants who had abrupt cessation of steroids (after 30 mg/day for 60 days) showed no evidence of adrenal difficulties. The search for the optimum dosage schedule deserves further study. Some aggressive hemangiomas will continue to enlarge while the infant is receiving 15 mg/day of Prednisone. We have not seen continued growth on doses of 20 or more mg/day.

Intralesional injections of Prednisone seem to have little advantage over systemic administration of the drug. Topical Betamethasone 17-valerate may be applied as a cream (for 6 hours daily) to capillary-cavernous hemangiomas with encouraging responses in very young infants when they show significant superficial dermal involvement.[21]

The original 7 infants in our 1967 report[23] have been followed for an additional 7 years (Figs. 3a to 3j). Although several have undergone later reconstructive surgery to remove residual noninvoluted vascular masses, or to correct deformities of features resulting from earlier



FIGS. 3h. Deep angioma was removed from the tip of the nose and the columella was constructed from dorsal nasal tip skin. The tumor in the upper lip has been removed and a mucous membrane flap is being inserted to replace the scarred vermillion. Two years later she has residual unresolved tumor in the left upper eyelid producing some degree of ptosis.

ulceration, none has had later recurrence of growth of hemangioma and none has developed late or persistent undersirable effects of the steroid therapy.

The mechanism of action of steroids on enlarging hemangiomas is still unclear, but Zweifach, Shorr, and Black[24] have shown that, in adrenalectomized rats, corticosteroids increased vascular sensitivity to circulating vasoconstrictive agents (Fig. 3e). Direct observations have also been made on the circulation in hamster cheek pouches with animals in adrenal insufficiency.[22] These investigators showed that intramuscular cortisone acetate produces arteriolar constriction and narrowing of the precapillary sphincters. In addition, they noted leukocyte coating of the endothelial walls. It seems not unlikely that rapidly proliferating blood vessels such as those seen in capillary cavernous juvenile hemangiomas may be highly susceptible to these mechanisms of steroid action.

Once the enlargement of a capillary cavernous hemangioma has been reversed and the family sees involution beginning, the surgeon and pediatrician will feel less pressure and more composure as they plan the future management of the remaining deformity and other symptoms (Figs. 4a and 4b). Areas on the face and neck that become ulcerated will usually leave sufficient scarring to require later plastic revision or skin grafting. A great variety of reconstructive procedures may be required for optimum correction of the resulting defects. In general it is wise to begin the major portions of this reconstruction before school age in order to minimize the adverse impacts of deformity on the child's self-image.

Our experiences with the use of CO_2 snow and liquid nitrogen to freeze capillary cavernous hemangiomas lead us to urge that these methods be abandoned. The surface scarring is hard to correct later and cold injury is relatively ineffective with the deeper portions of large lesions.

Irradiation Treatment of Hemangiomas

Gamma or beta irradiation therapy should never be used to treat any form of hemangioma. Indeed, Fost and Esterly[6] report "irradiation has been associated with damage to epiphyses, breasts, gonads, skin, lens and

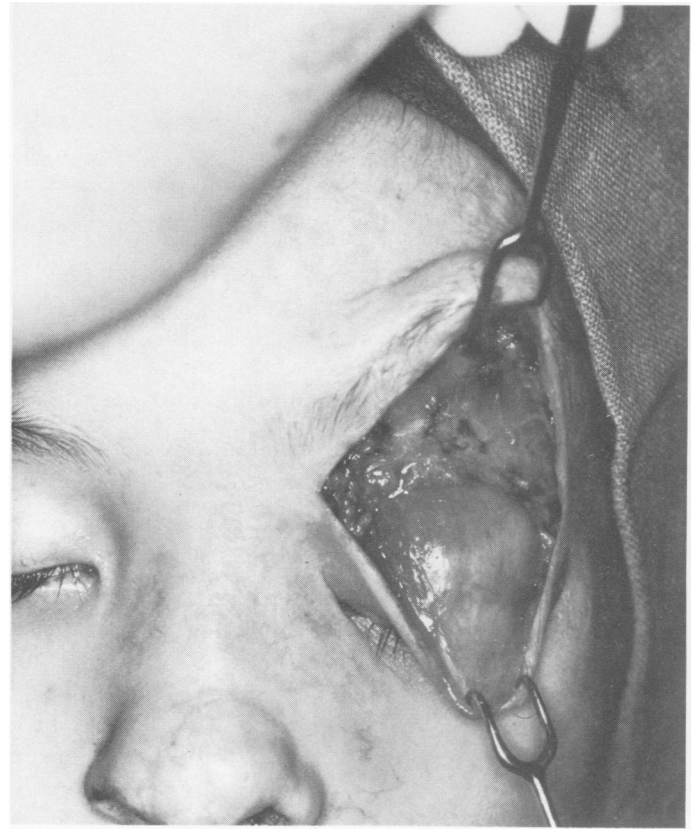


FIG. 3i. Massive deep angioma is still present in the left upper eyelid. At age 7 it was removed, taking care to preserve the adjacent levator palpebrae muscle fibers.

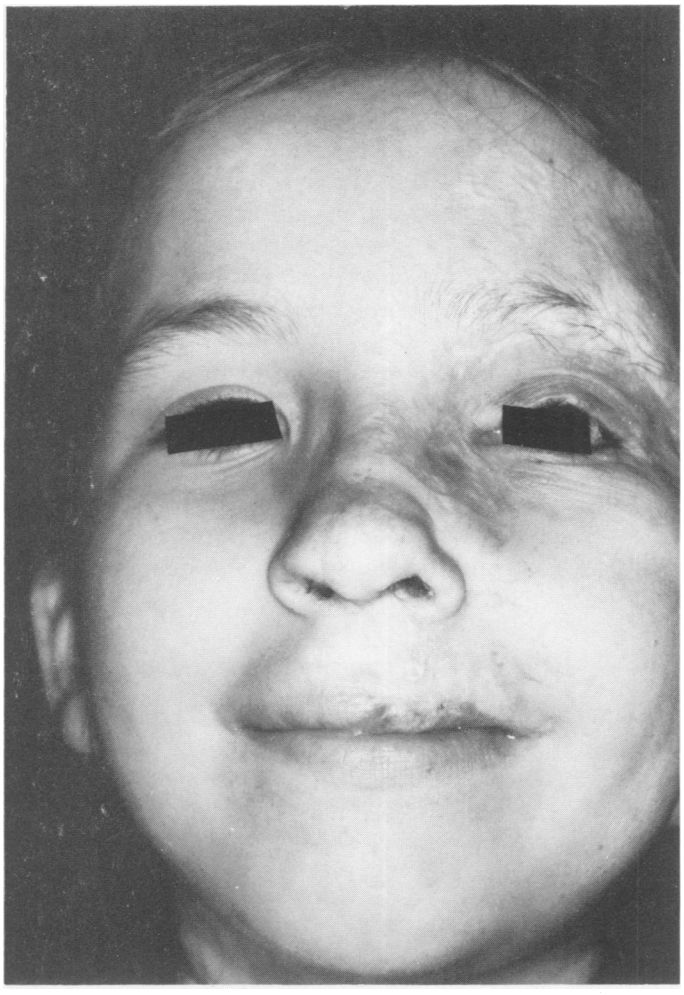


FIG. 3j. Present condition of the patient. She now needs only minor procedures to abrade and revise superficial scarring of nose, lip, and forehead.

thyroid, and in a large series yielded no better results than in untreated controls." They further point out that "the complications were ten times that in the untreated patients" (Figs. 5a and 5b).

There are numerous reports in the literature of carcinogenesis of the skin and deeper structures following the use of even light doses of gamma irradiation to treat hemangiomas in children (Figs. 6 and 7a–c). More recently Li, Cassady, and Barnett[13] have reported on 4,746 patients treated between 1946 and 1968 at the Boston Children's Medical Center by irradiation of hemangiomas during infancy. Despite the fact that the doses were all between 300 and 600 roentgens in air from a 50 cvp orthovoltage unit, there have already been three cancer deaths in this group with a mean followup of only 10 years. Two of these deaths were from lymphosarcoma and one from testicular teratoma (in this instance the irradiation included the testicle in the field of irradiation). Although the sample is still statistically small, one must be concerned about additional deaths from malignancy in this group of young patients in the years ahead. Li and Cassady[13] also point out "this was relatively low dosage x-ray and most (of the energy is) absorbed in the skin. Most induced skin cancers are curable and would not be ascertained by the methods of this study."

It is startling that in modern times a single hospital would continue to irradiate over 200 infants a year for a benign disease such as hemangioma!

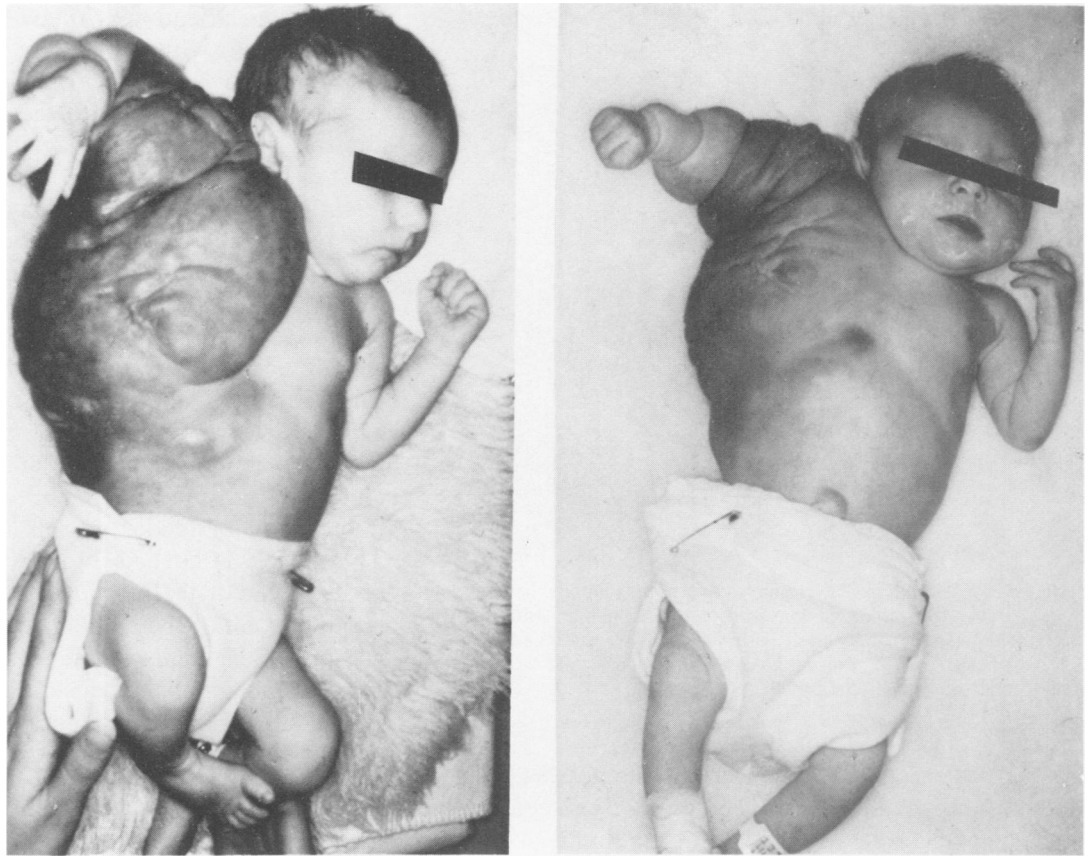
Disseminated Intravascular Coagulation (DIC) in Association with Giant Hemangiomas

Disseminated intravascular coagulation has been stated by Kwaan[11] and others to be "not a pathologic entity but a pathophysiologic intermediary mechanism of disease." This condition is associated occasionally with severe and uncontrollable bleeding in the postoperative period following surgical resection of portions of large hemangiomas. It seems probable that this condition is caused by hypoperfusion in a large hemangioma and that surgery may result in additional damage to the vascular endothelium of the remaining nonresected hemangioma. This leads to surface clotting, followed by the release of tissue thromboplastins into the circulation of the patient. Intravascular clotting then appears to produce microthrombi in innumerable small vessels throughout the body. Thrombi do not appear to form in the large blood vessels. The microthrombi cause a removal of platelets and fibrinogen from the circulation. Also factors II, V, VIII, X, and XII may be reduced in the circulating blood. As a result of these subtractions of elements important for clotting, bleeding may occur and may respond very poorly to treatment with fresh whole blood or platelet transfusions.

The fibrin clots formed in patients with DIC tend to cause the release of plasminogen activator from the endothelium in amounts 5 and 10 times above normal. This in turn causes a secondary fibrinolysis at a rate that exceeds the patient's ability to synthesize new fibrinogen. The breakdown products of this secondary fibrinogen degradation have large molecular weights (50–270,000) and at least 4 have been found to be effective antithrombin factors. These degradation products (FDP's) thus tend to further increase bleeding.

In DIC, the kidneys are commonly involved. Oliguria and hematuria may result from microthrombi forming in the glomerular capillaries. Patients may also have hemoptysis, adrenal failure, pancreatitis, or gastrointestional bleeding. Some patients with DIC will show hemolytic anemia and reduced plasma haptaglobins.

DIC may occur with a variety of disorders and all of these have the common denominator of being capable of producing procoagulants. They include such clinical con-



FIGS. 4a and b. (left), Massive rapidly growing capillary-cavernous hemangioma in newborn infant with associated thrombocytopenia. Bleeding problems and poor nutrition of the infant made the prospects of surgery formidable. (right) Ten days after the initiation of steroid therapy, the infant had shown this response. Further regression occurred over a period of months, making surgical excision of the residual masses a practical matter. Steroid therapy has proved most effective in the most massive lesions where effective treatment is most urgently needed.

ditions as massive red cell hemolysis, placental separation in pregnancy, massive burns, fat embolism, shock, pulmonary embolism, some allergies, as well as massive hemangiomas.

Jona[10] has reported the case of a 16-year-old boy with 19 hemangiomas, melena and anemia. After each attempt to resect one of these hemangiomas, he experienced severe disseminated intravascular coagulation and massive bleeding. He responded well to treatment with heparin (3000 international units q. 4 hours intravenously) and aspirin (300 mg orally q. 6 hours).

Heparin interferes with clotting and therefore decreases the tendency to further disseminated intravascular coagulation. Warfarin has not been found to be very effective in this condition. EACA (episilon-aminocaproic acid) may be helpful if fibrinolytic activity is excessive. It must be used with care as it may, however, cause serious thrombotic complications. When properly treated with heparin, patients with DIC should show a rise in fibrinogen and platelet levels within 24 hours. Surgical removal of the angioma should be completed while the patient is still on heparin.

Surgeons operating on any massive hemangioma must be on the lookout for the development of DIC and prepared to treat it appropriately. *Complete* resection of a hemangioma is less likely to initiate a bout of acute DIC than is *subtotal* removal of the endothelial mass.

Type 4: Arteriovenous Fistulae

Congenital arteriovenous fistulae differ from traumatic A-V fistulae in several important particulars. The communications are usually quite small in the congenital variety but they may be several hundred in number. Traumatic fistulae are usually one or two in number and may have larger diameters leading to the more typical type of cardiac enlargement and right-sided heart failure. *Congenital* A-V fistulae do not usually produce cardiac failure but may on occasion do so if the shunting is uncontrolled. There are two types of congenital A-V fistulae:

Arterial hemangiomas: These lesions are usually diagnosed clinically because of an elevated skin temperature, a rather bright red color (in some instances resembling a port wine stain), but with a steady tendency to progression and enlargement of the part. There is often a definite pulsation or thrill and bruit. Arterial hemangiomas may be apparent in very early childhood and enlarge at irregular rates until adult life. In females, the development of pregnancy may precipitate marked

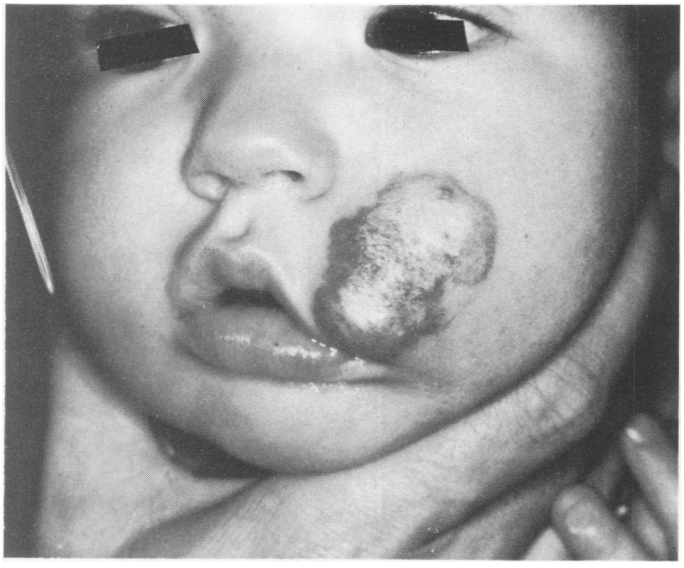


FIG. 5a. This child has a capillary-cavernous juvenile hemangioma of moderate size. Although the surface of the hemangioma showed some sclerosis, the deep mass within the cheek continued to grow. The physician treating this child recommended "mild" doses (800 rads total dose) of radiotherapy.

further enlargement with serious hemorrhage. Bone erosion and cartilage involvement may be seen. Arteriography will often reveal many tiny shunts within the local region.

Treatment: Peripheral ligation of vessels, sclerosing solutions, x-ray therapy, freezing, electrodessication, injection with boiling water, and many other modalities have all been tried unsuccessfully in efforts to control these alarming lesions.

About the only effective method to prevent progression of the vascularization is the *surgical resection of all of the A-V shunts* within these congenital lesions (Figs. 8a to c). When the fistulae are relatively limited in locality, this may be possible with an effective and permanent control. If, however, a significant number of shunts must be left behind, one can anticipate a slow, but relentless, reappearance of the warm pulsating mass with the return of abnormal hemodynamics leading to, in some instances, right heart failure.

In 1931 Brooks[2] described the use of small fragments of autogenous muscle for embolization of the arteries leading into congenital A-V fistulae. More recently, Luessenhop,[15] Cunningham and Paletta,[4] and Bennett and Zook[1] have reported further encouraging experiences with this technique. Small muscle fragments approximately ½ cm in diameter are inserted by arteriotomy into the major feeder vessels leading into the fistulae. The muscle is flushed into the distal artery with saline and x-rayed with the aid of radiopaque dye to determine location of the emboli, following which the vessels are ligated. Reduction in the size of the A-V hemangioma has been accomplished in some patients by this method.

Recently, Longacre, Benton, and Unterthiner[14] have employed the use of small silicone spheres as intravascular emboli in a patient with A-V fistulae. They report their patient still improving 4 months after insertion of 18 spheres between 1 and 3 mm in diameter into the distal stump of the external carotid artery.

We have used embolization as an adjunctive procedure

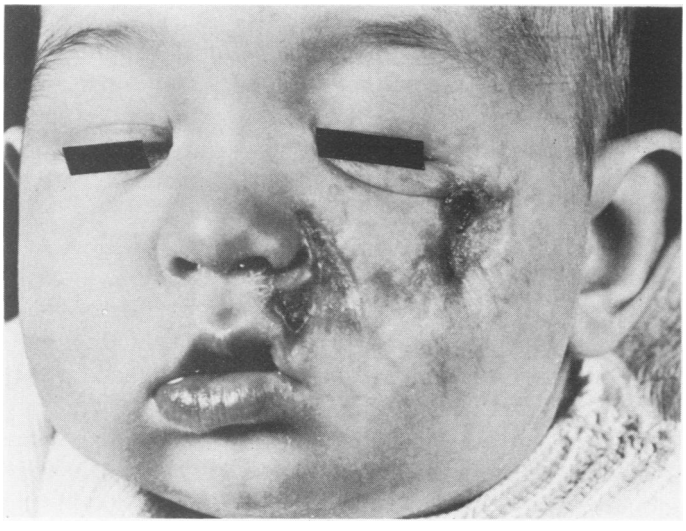


FIG. 5b. Although the hemangioma responded to gamma irradiation, the ulceration, scarring and deformity of the cheek 3 months later is awesome. Irradiation should never be used in the treatment of hemangiomas.

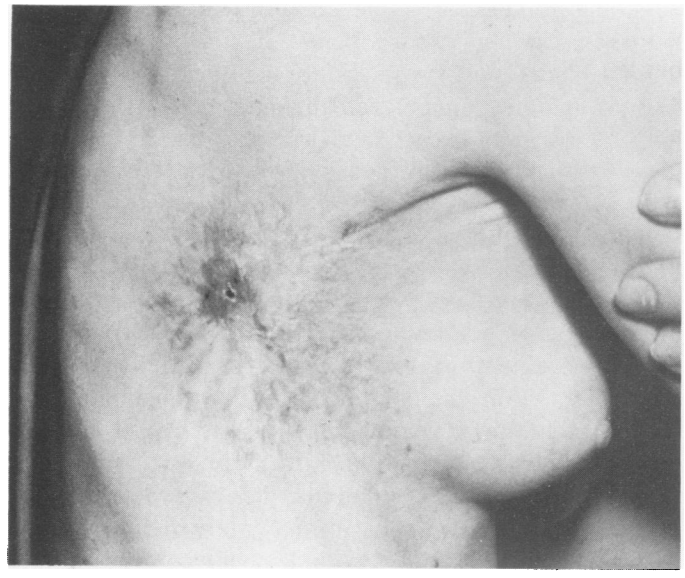


FIG. 6. Eighteen-year-old girl with a 1 cm capillary hemangioma of the chest wall treated in infancy by a 20-minute application of a radium plaque. The angioma disappeared but irradiation telangiectasia and scarring appeared within the skin. The ribs of the right hemithorax developed to less than one half the size of those on the left. The right breast is less than one half the size of the left breast, and a biopsy taken of the small ulcer shown revealed "squamous carcinoma." Many similar cases have been reported in the medical literature.

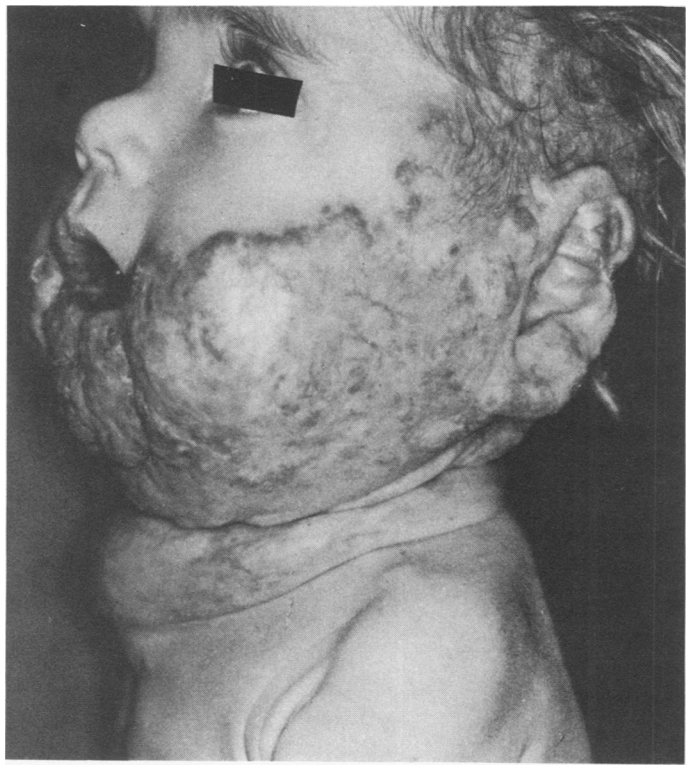


FIG. 7a. This 2-month-old infant with a massive capillary-cavernous hemangioma was losing weight, had severe stridor from laryngeal involvement, and had developed ulceration of the lips and hyperthermia. Irradiation therapy had been tried in desperation, but with minimal response. Emergency tracheostomy had to be performed directly through the bulky vascular tumor in the deep anterior neck. Intubation through the involved larynx seemed to hazardous.

with A-V fistulae in an effort to reduce size and minimize bleeding at the time of surgical resection. The use of hypotensive anesthesia and preliminary control of major arterial trunks have added to the safety and effectiveness of surgical control of the congenital A-V fistulae.

Hemangiomatous giantism: In some instances, congenital A-V fistulae seem to be associated with marked regional giantism of the involved part. Most often this involves an extremity but may involve the lower jaw, lip, or some other facial feature. There is usually a purple discoloration of the skin with marked enlargement of the underlying bones and soft parts. X-ray may reveal radiolucent areas in the shafts of long bones where these shunts exist. Bleeding is not uncommon and, indeed, ischemic ulceration may occur in an extremity as a result of the inefficient perfusion of distal parts of that extremity in the presence of multiple large shunts, more proximally located. Attempts to control enlargement or bleeding by proximal arterial ligation will usually lead only to further distal ischemia with pain and gangrene often developing. In the presence of cardiac enlargement, amputation of the part may be the only practical treatment.

Type 5: Cirsoid Angioma (Racemose Aneurysm)

A relatively uncommon form of hemangioma may be composed of large wormlike clusters of venous tributaries with purple discoloration of the overlying skin and a tendency of the lesions to become tense and swollen when the venous outflow to the region is obstructed. These may be seated quite deeply and at times the walls of these angiomas will contain many fine cutaneous nerves that may produce significant pain when the lesions become engorged.

Cirsoid angiomas may enlarge slowly. They do not involute and respond only to surgical excision. Usually staged procedures in combination with appropriate reconstruction will produce the maximum relief.

Lymphangiomas

Although this paper will not deal exhaustively with the treatment of lymphangiomas, it is essential that several important distinctions be made between hemangiomas and lymphangiomas in regard to clinical manage-

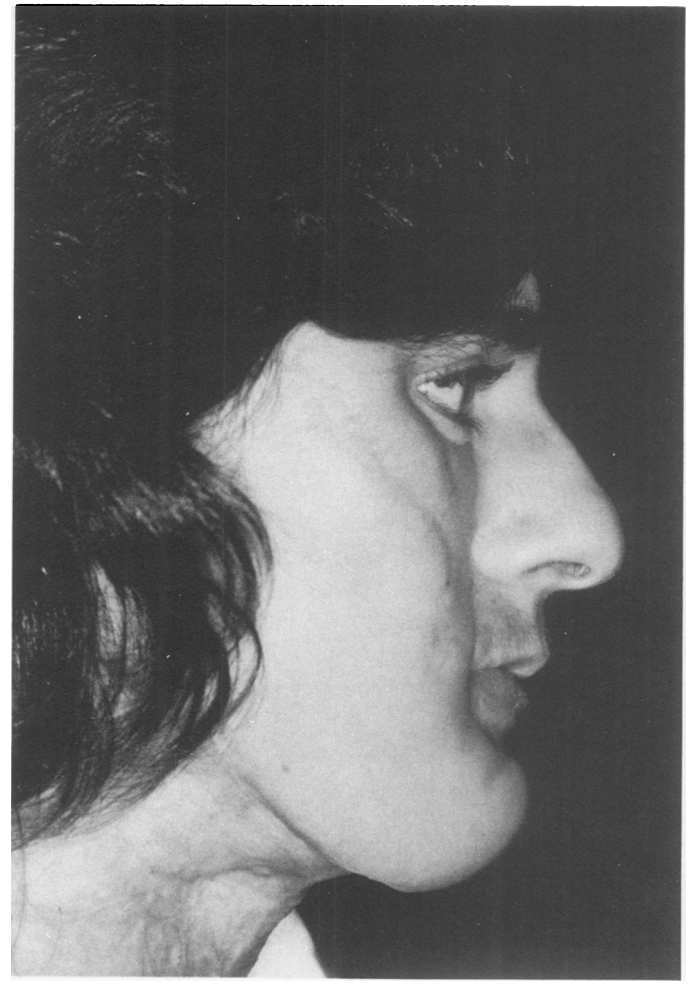


FIG. 7b. Bone graft has been supplied to elevate the right eye and flap has been fitted into parotid region. Chin pad is unduly prominent.

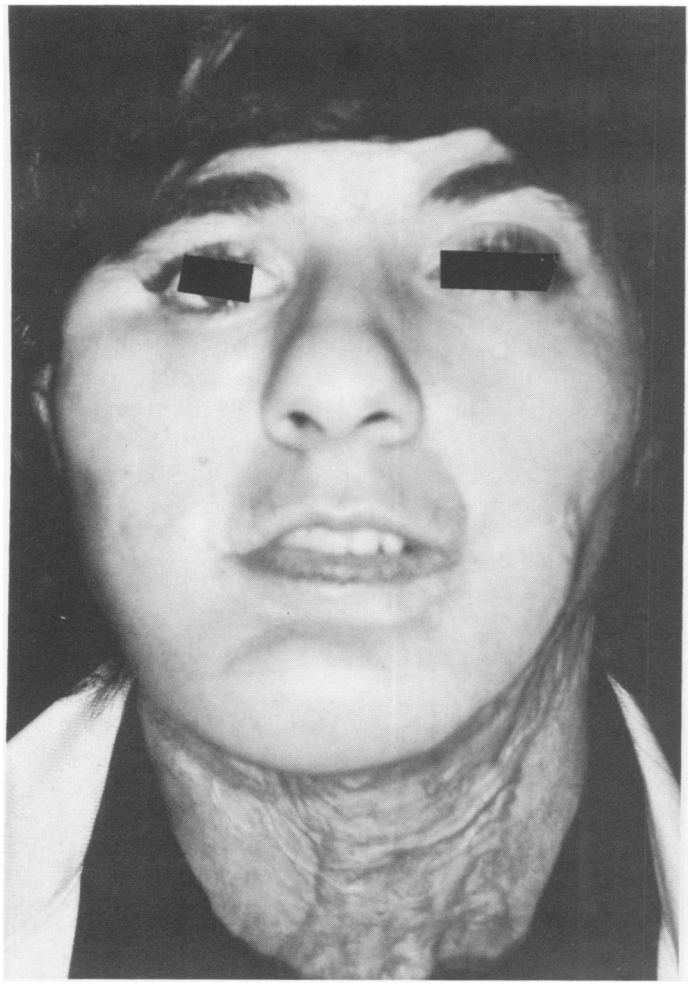


FIG. 7c. Present condition shows facial reconstruction largely completed and now awaiting a cosmetic overlay graft to provide a smoother surface texture to the grafted anterior neck. Much of the reconstruction over the years in this young woman was required to repair the effects of gamma irradiation on bone and soft tissue.

ment. *Contrary to widespread medical opinion, lymphangiomas and hemangiomas are seldom intermingled in the same patient.* Although some lymphangiomas do have a dark red color suggestive of hemangioma, this is due to different pathology, involving blood pigment in stasis within lymphatic channels, unlike the red tones produced by the rapidly circulating blood in the capillaries and larger vessels of a hemangioma.

Like hemangiomas, lymphangiomas may be diffuse or cystic in nature and, in the absence of obvious blood pigment within the sinusoids, it is difficult for a pathologist to distinguish the endothelium of these two lesions. Despite this, they are entirely different from the standpoint of prognosis, clinical symptoms, and methods of treatment.

Lymphangiomas may be found in tissues quite deep to the skin. Very commonly, in the head and neck region, they also involve the skin, producing a light brownish pigmentation, frequently small keratotic papillomas and occasionally drainage of clear yellow lymph from one of these skin excrescences. When the lymphangioma reaches the surface of any body cavity that is lined with mucous membrane or serosa, it produces multiple tiny vascular tufts that are pale in color, and each with a single pink center. This gives mucous membrane a typical appearance that is pathognomic for the diagnosis of lymphangioma.

At times, lymphangiomatous enlargement of soft tissues may be accomplished by localized giantism of that part of the body.[5] This symmetrical enlargement of the part may involve soft tissue, bone, teeth, or indeed any structure. The pattern of enlargement often reflects the peripheral sensory nerve distribution to that region. In many, but not all, instances of localized giantism, the careful examination of histologic sections will reveal plexiform neurofibromatosis to be a part of the local picture.

When left untreated, the natural history of the localized lymphangioma is usually one of very slow enlargment, accompanied after a period of months or years by the tendency to recurrent episodes of cellulitis and ascending lymphangitis. This may become more frequent with the passage of time producing episodes of severe systemic illness and fever. These bouts of infection leave the lymphangioma increasingly indurated. When a lymphangioma is located in the region of the oral cavity or neck of an infant, respiratory obstruction is not uncommon and may even be fatal. In the facial region,

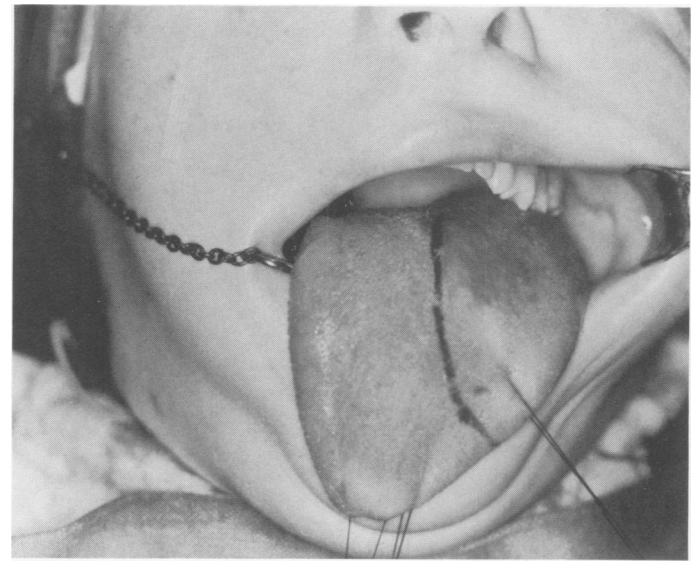


FIG. 8a. This 16-year-old girl had enlarging pulsating, warm, congenital A-V fistulae involving the base of the left tongue. Alarming, bleeding had occurred on two occasions. The lesion was first noted approximately 3 years before.

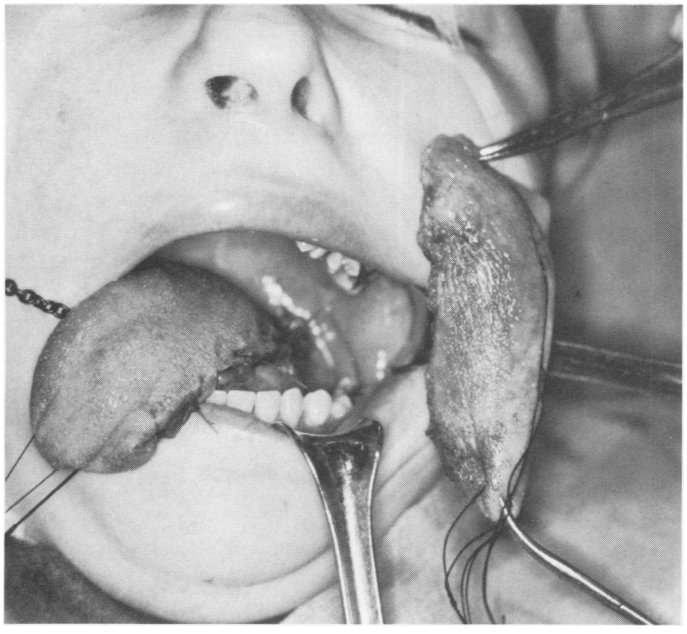


FIG. 8a. Arteriograms shows the shunting confined to the left half of the tongue. A surgical resection of one third of the tongue was carried out intraorally, after obtaining control of the carotid arteries. A pharyngeal flap was used to close the base of the tongue.

deformity often brings the patient to the attention of the plastic surgeon. Lymphangiomas do not spontaneously involuate in the manner of juvenile capillary hemangiomas but the tendency to growth becomes less noticeable after about 17 or 18 years of age.

Treatment: The lymphangioma is almost totally resistent to any form of irradiation of cryotherapy. Although sclerosing agents have been used to reduce the size of cystic hygromas, it is a relatively unsatisfactory method of management and greatly complicates any later surgery that might be required to reduce the mass.

In the vast majority of instances, surgical excision is the only effective method of controlling the lymphangioma. Since the involved tissue very rarely becomes malignant, it is usually safe to retain some portions of the affected area for use in local reconstruction. It is not essential that the entire lymphangioma be removed in order to control or prevent further symptoms from developing. When localized giantism is present with a lymphangioma, we prefer to perform longitudinal reductions of the massively thickened sensory nerves supplying the enlarged region. This is done at the earliest practical date in the hopes of further reducing stimuli to overgrowth of the part.

The patient with a massive lymphangioma presents one of the most difficult and complex problems seen by plastic surgeons and may require an extensive gamut of reconstructive techniques to eliminate deformity and restore function.

General Principles in the Treatment of Angiomas

The most valuable methods currently of value in the treatment of the various clinical varieties of hemangioma include the use of: 1) *Observation* to await involution in the case of neonatal staining or the smaller circumscribed juvenile strawberry capillary hemangiomas. 2) "*Heavy dose–short course*" steroids (or occasionally sclerosing agents) to accelerate involution is the principle treatment approach to growing juvenile capillary, or capillary-cavernous hemangiomas. 3) *Intermittent compression* of a hemangioma is of value when there is associated thrombocytopenia. Subsequent surgical excision of the tumors with plastic closure may often be required. 4) *Surgical excision in combination with skin grafts or pedicle flap repair* remain the most valuable treatment approaches for intradermal capillary hemangiomas, arteriovenous fistulae, or cirsoid hemangiomas. 5) *Medical tattooing and cosmetic cover-up creams* of port wine stains have both been disappointing. Skin graft replace-

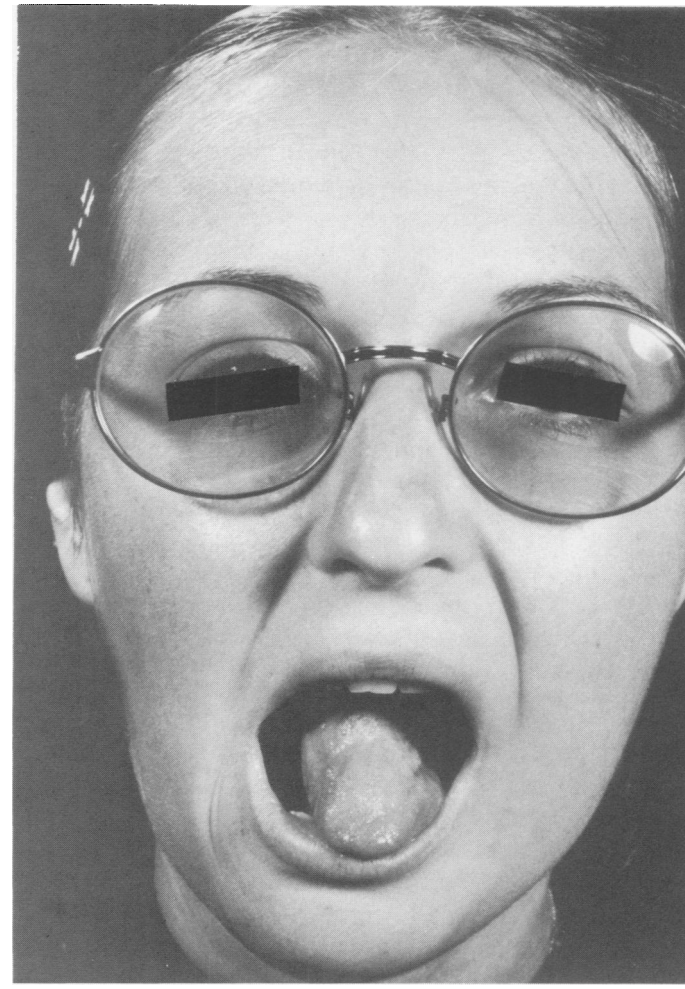


FIG. 8c. Appearance of tongue two years later with some tipping of the base of the tongue. Excellent speech and no problem in swallowing.

ment of port wine stains is becoming more effective with better graft techniques and the use of skin from donor sites with "blush" capabilities. 6) There is very little indication for the use of CO_2 snow, liquid nitrogen, or electrodessication. 7) Any form of irradiation is always contraindicated in the treatment of hemangiomas! 8) Lymphangiomas usually require staged surgical resection and reconstructions. They do not involute and portions of tissue containing the abnormal lymphatic network may be used in the reconstructions.

Comment

Hemangiomas have been defined and a classification has been proposed which combines the essential features of prognosis and optimum treatment methods. Treatment of each major variety of hemangioma has been outlined and special emphasis has been placed on the use of systemic steroid therapy as an invaluable aid in the management of the massive juvenile capillary-cavernous hemangiomas of infancy.

Some consideration has been given to the surgical treatment of port wine stains and other intradermal hemangiomas. Current approaches to the management of congenital arteriovenous fistulae are outlined.

Major complications of gamma and beta irradiation therapy are demonstrated to support the position that they are unwise and contraindicated methods of treatment for all forms of hemangiomas.

More attention should be paid to the psychic impacts of deformities resulting from hemangiomas. In that regard, plastic surgical techniques can be used more effectively and applied at younger ages than has been the custom in the past.

Several important distinctions have been made between lymphangiomas and hemangiomas in regard to prognosis and treatment.

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Discussion

Dr. Stephen R. Lewis (Galveston): Dr. Edgerton does not like Covermark for covering the port wine stain. I'm not sure that his skin grafts are that much prettier than a port wine stain covered with good cosmetics. We have found that the adult male, or most males, will not use Covermark, but females use it, and use it relatively well.

Our skin grafts usually are not as mobile as that skin that he has removed, and I think our end result with cosmetics is usually a little bit better than our skin grafts, although we have skin-grafted a number.

So many times when we have excised the port wine stain, we see a recurrence of the lesion at the border when we're well around the primary lesion. I wonder if Dr. Edgerton has had this same complication or problem with his grafts.