

HEREDITARY HEMORRHAGIC TELANGIECTASIA

WITH REPORT OF THREE FAMILIES AND A REVIEW OF THOSE
PREVIOUSLY RECORDED *

WALTER R. STEINER, M.D.
Visiting Physician, the Hartford Hospital
HARTFORD, CONN.

In 1865 Babbington¹ reported an interesting family in which recurring epistaxis had been observed for five generations. The transmission was through both male and female, but no mention was made of telangiectases in any of them. The first family in which these telangiectases were found was reported eleven years later by Legg,² who described numerous small nevi on the face, forehead and trunk of his patient. They developed about his 41st year. These cases were followed by others reported by Chiari,³ Rendu,⁴ Osler,⁵ Josserrand,⁶ Kelly,⁷ Hawthorne,⁸ Parkes-Weber,⁹ Phillips,¹⁰ Waggett¹¹ and Ballantyne.¹² Rendu, however, was the first of these observers to associate the presence of epistaxis with multiple telangiectases as clinical manifestations of a distinct morbid entity. Henceforth the erroneous diagnoses of hemophilia, acquired angiomas from cirrhosis of the liver and hemorrhagic diathesis were no longer applied to this condition. Further interest in its study was aroused by the papers of Sir William Osler, whose first published article on it appeared in 1901. Most of these cases were carefully abstracted in 1909 by Hanes¹³ when he reviewed the literature on this subject and accurately described two

*Read in part at the meeting of the American Climatological and Clinical Association, Washington, D. C., May 9, 1916.

1. Babbington: *Lancet*, London, 1865, **2**, 362. Recently Lane (*Jour. of Heredity*, 1916, **7**, 132) has described hereditary nosebleed observed in a family for three generations.

2. Legg: *Lancet*, London, 1876, **2**, 856.

3. Chiari: *Erfahrungen auf dem Gebiete der Hals und Nasenkrankheiten*, Vienna, 1887, p. 60.

4. Rendu: *Gaz. d. hôp.*, 1896, **49**, 1322.

5. Osler: *Bull. Johns Hopkins Hosp.*, 1901, **12**, 333.

6. Josserrand: *Bull. de la Soc. méd. d. hôp. de Lyon*, 1902, **1**, 244.

7. Kelly: *Glasgow Med. Jour.*, 1906, **65**, 411.

8. Hawthorne: *Lancet*, London, 1906, **1**, 90.

9. Parkes-Weber: *Lancet*, London, 1907, **2**, 160.

10. Phillips: *Proc. Roy. Soc. Med.*, 1908, **1**, Laryngological Section, p. 45.

11. Waggett: *Proc. Roy. Soc. Med.*, 1908, **1**, Laryngological Section, p. 70.

12. Ballantyne: *Glasgow Med. Jour.*, 1908, **70**, 256.

13. Hanes: *Bull. Johns Hopkins Hosp.*, 1909, **20**, 63.

additional families, so that fifteen were then reported.¹⁴ Since then eleven more families have been described, including the three here detailed, so that this condition has now been found to exist in twenty-eight families.¹⁵ Other, isolated, instances, with no further cases in any of their families, have been reported by Chauffard,¹⁶ Kelly,¹⁷ Fox,¹⁸ Galloway,¹⁹ Lack²⁰ and Adamson,²¹ but I have omitted all consideration of them on account of their lack of any hereditary tendency toward this condition.

It has been defined as a hereditary affection, manifesting itself in localized dilatations of capillaries and venules, forming distinct groups or telangiectases, which occur especially on the skin of the face, nasal and buccal mucous membranes and give rise to profuse hemorrhage, either spontaneously or as the result of trauma (Hanes).

In its causation, a hereditary tendency is the only factor which is constantly present, although others, such as syphilis, alcohol and traumatism, have been irregularly reported. A history of syphilis, however, is only recorded in three instances, yet its unrecognized presence may probably have existed in a somewhat greater frequency. Stokes²² recently has written an interesting article showing that syphilis, chronic plumbism, hyperthyroidism and nephritis may cause cardiovascular degenerative conditions which result in the formation of generalized telangiectasia. Alcohol, likewise, is a most inconstant factor, which is only thrice stated to have been present, and traumatism, of etiologic importance, is just as rarely seen, for it appears but two times in the histories of the twenty-eight families exhibiting this condition, although

14. In this enumeration the family reported by Josserand was unintentionally omitted and Osler's second case seemed to be an isolated instance, not of the family type. Later another member of the family, similarly affected, was discovered. Gottheil's family was also omitted from Hanes' list. It was published shortly before Hanes' article appeared. Hanes' list includes Babbington's family, in which epistaxis was observed for five generations, but no mention is made of telangiectases in any of its members. It probably should be added to this group, but, on account of the absence of positive evidence, I have omitted it.

15. Laffont: *Presse méd.*, 1909, **17**, 763. Langmead: *Proc. Roy. Soc. Med.*, 1910, **3**, Clinical Section, p. 109. Audry: *Rev. de méd.*, 1911, **30**, 22. Osler: *Riforma med.*, 1911, **27**, 57. Van Wagenen: *Med. Rec.*, New York, 1912, **81**, 109. Sequeira: *Proc. Roy. Soc. Med.*, 1912-1913, **6**, Dermatologic Section, p. 128. Gjessing: *Hospitalstidende*, 1915, **8**, 1151. Hutchison and Oliver: *Brit. Med. Jour.*, 1916, **9**, 67.

16. Chauffard: *Bull. et mém. Soc. méd. d. hôp. de Paris*, 1896, **13**, Series 3, p. 352.

17. Kelly: *Proc. Roy. Soc. Med.*, 1908, **1**, Laryngological Section, p. 45.

18. Fox: *Brit. Jour. Dermat.*, 1908, **20**, 145.

19. Galloway: *Proc. Roy. Soc. Med.*, 1910-1911, **4**, Clinical Section, p. 42.

20. Lack: *Jour. Laryngol.*, 1909, **24**, 185.

21. Adamson: *Brit. Jour. Dermat.*, 1909, **21**, 219.

22. Stokes: *Am. Jour. Med. Sc.*, 1915, **149**, 669.

it must be stated, as Hanes has previously shown, that the telangiectases are generally located at points most subject to frequent slight traumatisms. The bleeding in these cases is often so severe that a condition of marked anemia is induced, which may be an important factor in the development of the telangiectases. Certain it is that the bleeding generally precedes the formation of these localized dilatations of the capillaries and venules, and may do so with many years intervening between them. In some of the cases, also, a respite from the bleeding appears to be coincident with the disappearance of the telangiectases. Cases have been reported in England, France, Germany, Denmark and the United States. The Anglo-Germanic race has furnished most of the examples, followed by the Latin and Scandinavian.

Kelly thinks that October to May, inclusive, are the months in which the bleeding is most apt to occur, but other observers have not corroborated this fact. Hutchison and Oliver thought their patient bled more during the summer months. It has been noted in ninety-two males and seventy-three females, so its distribution among the sexes is probably about equal. Both sexes are able to transmit it. In the published cases thirty-two women have transmitted the tendency as against thirty-five men. The pathology of this condition has been investigated on three occasions. The first case was when Dr. Mabel Austin examined at Dr. Osler's suggestion a section of the nasal mucous membrane of a patient, in the first family which Dr. Osler reported. This patient was the first to come to necropsy, which revealed "cancer of the stomach, mesentery and omentum, liver, retroperitoneal glands, lungs and brain." In the stomach there were also found "a dozen round foci, each from 3 mm. to 4 mm. in size, which at first looked like ecchymoses, but were dilated venules and capillaries." The sections of the septum of the nose showed large dilated veins just beneath the epithelium. Our information from the second case comes from a biopsy which Dr. Hanes performed on one of his patients. The tissue was removed from a telangiectatic spot and showed "the obliteration of the papillae of the corium, together with the absence of the usual undulations of the stratum germinativum. This is doubtless due to the relatively enormous dilatation of the blood vessels of the corium, which are seen as wide spaces, lined by a single layer of endothelium lying immediately subjacent to the greatly attenuated epidermis. These dilated vessels can be traced well down into the subcutaneous fatty tissue. A study of sections stained by various special methods failed to reveal any muscular or elastic tissue in the walls of the dilated superficial vessels, although the less dilated vessels in the subcutaneous fatty tissue show the normal arrangement of the tunicae." Consequently, Hanes refers to the insufficient protection of the dilated vessels and

states that it is not surprising that trivial traumatism produce marked hemorrhage.

In one member of the family reported by Gjessing a microscopic examination was made from an angioma on the left cheek. In the corium blood-filled cavities were found, surrounded by a single layer of endothelium and a thin stratum of connective tissue, devoid of elastic or smooth muscle fibers. In some places the cavities extend upward to the epidermis, where the papillae were flattened. In one spot the cavity extended down into the subcutaneous tissue, where, in the surrounding area, numerous well-developed hair follicles and sebaceous glands were seen. In this situation the elastic fibers were as numerous and well developed as in normal tissue. The changes then consist in a dilatation of the capillaries of the small veins, with the formation of telangiectases of the three types which I shall later describe. These dilated vessels, being only lined by endothelium without the additional presence of elastic or muscle fibers, become consequently very liable to rupture, induced either spontaneously or by trauma.

The chief symptom is hemorrhage, which may come from the telangiectases in the nose as an epistaxis or from those situated elsewhere. These locations are the conjunctivae of the upper and lower lids, the ears, cheeks, nose, lips, mucous membranes of the mouth in the region of the hard or soft palate, the uvula, buccal mucous membranes or gums, tongue, neck, trunk, back, arms, finger tips, under the nails, or on the feet. The bleeding varies greatly in frequency. It may occur three to four times daily, once or twice a week or even with a greater interval of freedom. Kelly has described a patient in whom it was most apt to take place between the months of October and March, and would be both profuse and frequent. The amount of blood lost also varies greatly. In Kelly's patient, just referred to, it was so marked that death from syncope finally resulted. Legg and Chiari have reported cases in which dropsy ensuing from loss of blood caused a fatal termination, while Phillips has recorded a case in which death came from hemorrhage of the gums. Chiari also mentions a child, in one of his families, who died from a severe nosebleed, and Gottheil speaks of a male dying from the same cause.

The bleeding may be a slight trickle from the nose or the spots, or come with more force. One of Hanes' patients found it not uncommon for him to injure one of the lingual telangiectases while eating, so that the blood would actually spurt from the injured spot and render further progress with the meal impossible. Gottheil has described two varieties of bleeding in one of his cases. From the nose it occurred as a slow trickling, lasting perhaps ten or fifteen minutes, while from the visible lesions of the tongue and lips it came as a sudden projectile spurt,

sometimes reaching out a foot or two, if on the lips, or if his mouth was open, and stopping spontaneously in two or three minutes. Langmaid speaks of his patient as suffering for about twenty years from frequent epistaxis and adds that occasionally his face or tongue has burst out bleeding. Legg also speaks of the bleeding from the spots as being frequently spontaneous in origin in his case, but generally in all the instances it is the result of traumatism, while the hemorrhage from the nose is spontaneous in its onset.

Less severe bleeding has produced, by the resulting anemia, vertigo, headaches, weakness, dyspnea on exertion, palpitation and swelling of the ankles. In one instance fainting was induced after a prolonged epistaxis, while another patient, depleted from the hemorrhages, finally came down with an endocarditis. The prostration from this cause prevented another from working for about three years. The onset of the epistaxis is generally in early childhood, but the attacks become more severe and prolonged as the patients advance in years, the period between the 35th to 38th years being the time when the increase is especially noted. The telangiectases seen in this condition are of three varieties: the pinpoint, which is most apt to be seen on the skin of the hands and face, and which may be readily overlooked; the spider form, which is the most common and which Parkes-Weber prefers to call spider angiomas rather than spider nevi; and the nodular type, which may originate in the center of a spider angioma and finally form a solid vascular tumor, split pea in size. They are most frequently seen on the nasal and buccal mucous membranes and on the mucocutaneous junction of the lips, but may be found in the other locations mentioned above. They begin as capillary dilatations and are bright red in color. Later the venules give the cutaneous telangiectases a violaceous or purple color by participating in their formation. The spots on the mucous membranes, however, always remain a bright red. They may be seen early in life, for Hanes has observed them in a boy of 8 years, but generally, if present at such an age, they are not especially numerous, for they do not attain their full number until after the age of 35. Even then they appear and disappear with marked frequency and seem to bear some relation to the bleeding, being less marked if a considerable respite from hemorrhages is observed. Parkes-Weber speaks of a vicious circle being established by the repeated attacks of bleeding giving rise to a grave condition of anemia, which in its turn increases the tendency to hemorrhage.

In Parkes-Weber's case a small patch of chorioretinitis was found in an examination of the fundus of the right eye by the ophthalmoscope, while in Sequeira's case the retinal vessels were thickened, but there were no hemorrhages.

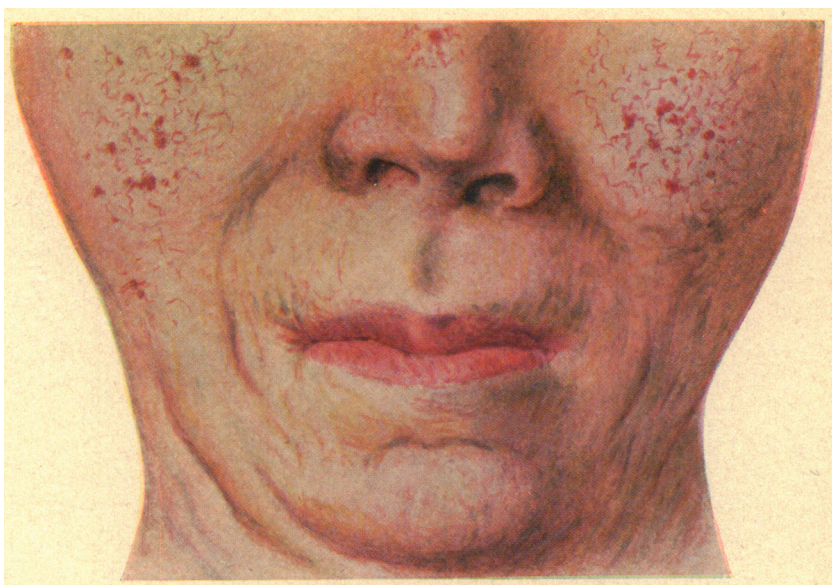


Fig. 1.—Telangiectases on the face. The family tree of this patient is given in Figure 3 (Family 28, Steiner 1, III, 22).

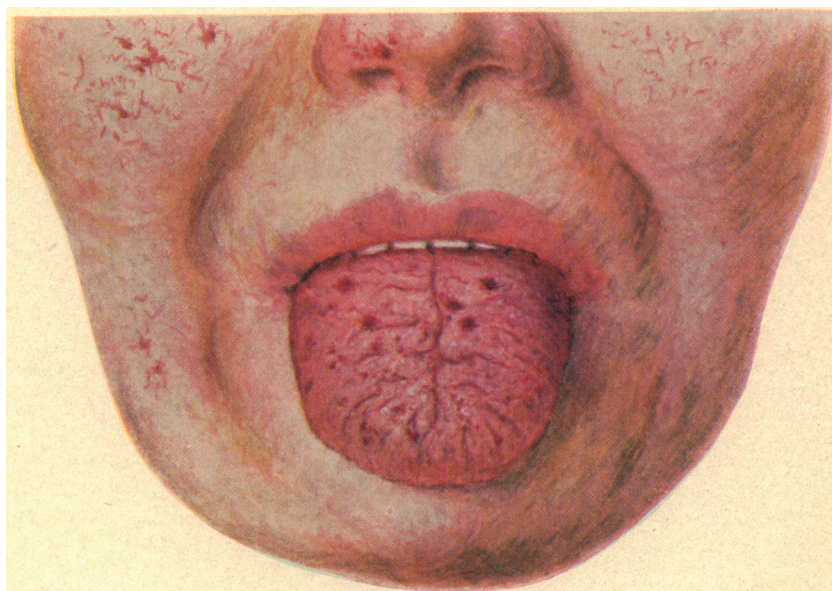


Fig. 2.—Telangiectases on the tongue. The family tree of this patient is given in Figure 3 (Family 26, Steiner 1, IV, 12).

Before Rendu's time the disease was generally considered hemophilia. Chiari, however, caught a glimmer of the real truth when he first diagnosed the condition as one of telangiectases of the mucous membranes, which were hereditary in a family. Unfortunately, after a further study of his cases, he regarded the affection as due to hemophilia. The epistaxis, or the telangiectases, may exist alone in certain members of the families implicated, but the hereditary tendency in them and the finding of the cardinal symptoms combined in other members of the family will make the diagnosis simple. The fact that the females are not affected by hemophilia may be, at times, of diagnostic value.

The outlook is generally not very promising, for the hemorrhages are apt to increase in severity as middle life is reached, when the telangiectases are frequently first seen. In one of the cases, however, there was a cessation of the bleeding as this period was approached, while another bled only after this time from her nose and the telangiectases elsewhere, at each menstrual period. In four out of the 191 cases death has been directly attributed to the hemorrhages, in others it has involved much invalidism, while a third and smaller group has not been much incapacitated by them.

Rendu used with excellent result, in his case, an application on the nasal mucous membrane of a powder of antipyrin, tannin and powdered sugar, and Coe has reported on the beneficial employment of calcium lactate and iron in his case, which he published as one of hemophilia. It is hard, however, to see the rationale of this treatment, for the disease is due to a developmental defect in the blood vessels and measures to increase the coagulability of the blood ought not to yield good results. The use of iron and the application of the thermocautery to many of the spots might more reasonably explain the patients' improvement. A bead of chromic acid, fused on a probe, was used with excellent success by Hanes, who recommends the method highly, as the action of the caustic may be checked at any time by the application of an alkali. The repeated use of this method may be necessary to check the bleeding. After the cause is thus removed, the use of iron and arsenic for the treatment of anemia is indicated. Although it has been unsuccessful in some cases, the cauterization of the troublesome bleeding areas appears to be the most satisfactory form of treatment. The use of radium for the nodular telangiectases might also be of avail.

Since my interest was aroused in this condition from a case on the services of Sir William Osler, at the Johns Hopkins Hospital, in 1899, I have seen instances of it in three families and can give the following partial rehearsal of their histories.

FAMILY 26 (Figs. 1, 2 and 3, Steiner 1, 1916).—Recurring hemorrhages occurred in five generations and twenty-one members of one family; telangiectases were observed in five of them.

F. G., aged 42 years (Hospital No. 66559), was admitted to the Hartford Hospital Feb. 23, 1913, complaining of weakness, shortness of breath and frequent nosebleed (Fig. 3, III, 4).

Family History: The patient's grandfather (Fig. 3, I, 1) was a bleeder, as well as his father (II, 1) and one aunt (II, 6) all of whom are now dead. Of the aunt's family of ten children, two sons (III, 12 and 14) and two daughters (III, 22 and 24) are bleeders. One of the daughters (III, 22, Figs. 1 and 3) has a girl (IV, 18) who is also a bleeder, and one son (III, 12) has two daughters (IV, 10 and 12) and a son (IV, 17) who bleed likewise. One of these daughters, both of whom are married, has a daughter (V, 6) who bleeds from the nose. The histories of III, 22 and IV, 12 (Figs. 1, 2 and 3) will be given later. Another aunt (II, 8) has two sons (III, 29 and 30) who are subject to epistaxis, and an uncle (II, 3) has likewise two sons (III, 6 and 7) who suffer from the same difficulty. The patient (III, 4) has three sons and a daughter (IV, 1, 2, 3 and 4) who are similarly affected.

Present Illness: The patient was in excellent health until he was 14 years old, when he began to suffer from frequent attacks of epistaxis, which were exceedingly difficult to control. They recurred weekly or at more frequent intervals. When he was 16 years old, he had a slight hemorrhage from a small red spot on his face, which was readily checked. Three years later he entered the Prussian army and suffered frequently from the symptoms mentioned above, although the amount of blood lost was never very great. There was never any additional hemorrhage noted from his mouth or rectum, and his urine never contained blood. He remained in a stationary condition during the four years he served in the army and in this period was observed by army physicians and others who told him the bleeding could not be checked. At the expiration of his army service he emigrated to the United States, where he was frequently in very straightened circumstances. Occasionally he found employment in the Hartford coal yards. After two years he returned to Germany again, where he labored successfully for thirteen years as a grocer and lumberman. Then meeting reverses, he came to this country again, four years ago, and has since worked as a pedler. During all this time he has had frequent attacks of epistaxis and has gradually grown weaker. For the past year dyspnea on exertion as well as palpitation have been troublesome symptoms. The hemorrhages generally occur in the mornings and of late have been observed daily. His appetite has been good. There have been no digestive disturbances. His bowels have been constipated, but have recently been moved daily by medication. Of late years he has suffered from hemorrhoids, which occasionally bleed somewhat. He had measles when he was a child. He denies venereal disease. He has been a moderate smoker and drinker.

Physical Examination: The patient was a well-nourished, well-developed Jew. The lungs were negative on examination, but the heart, which was not enlarged, revealed a soft systolic murmur at the apex, which was not transmitted outward, but was heard with increasing intensity on passing upward, being loudest at the base, in the pulmonic area, where the second sound was slightly accentuated. Over both cheeks, on either side of the nose, numerous angiomas are seen, varying in size from pinpoint to one which measures 3 by 4 mm. in size, located 20 mm. below a midline drawn perpendicularly from the left lower eyelid. Two pinhead angiomas are also seen on the nose, and on the mucous membrane of the lower lip ten more are noted from pinpoint to pinhead in size. On the tip of the tongue, two are also observed, the larger measuring 2 by 2 mm., while the smaller resembles a pinhead. Back of the wisdom tooth, on the right lower jaw, there is also one of a similar size, as well as on the buccal mucous membranes and on the posterior aspect of the tongue. Two small ones are likewise seen on the anterior aspect of the helix of the

right ear, while six more are visible on the posterior surface. On the septum of the nose, as well as on the left inferior turbinate, similar angiomas are observed and some of them appear to have bled very recently. The mucous membranes of the nose as well as the conjunctivae are very pale in color.

Blood examination showed red blood corpuscles 4,696,000; leukocytes 4,800; hemoglobin (Dare) 60 per cent.; coagulation time three and a half minutes (Biffi Brook's instrument). A differential count of 300 leukocytes gave polymorphonuclears, 61 per cent.; lymphocytes, 26 per cent.; large mononuclears and transitionals, 12 per cent., and eosinophils, 1 per cent. There were no myeloblasts, no normoblasts or megaloblasts, no poikilocytosis, and but slight anisocytosis. The urine examination was negative. On March 5 Dr. E. Terry Smith examined the patient's nose and observed the presence of an old atrophic rhinitis. He also noted some telangiectases on the septum and left inferior turbinate, which he cauterized on two separate occasions. His other treatment consisted in the use of rest, iron and arsenic. Three days before discharge the patient's hemoglobin was 70 per cent., but the blood examination otherwise showed no change. On April 19 he was seen with me by Sir William Osler, who confirmed the diagnosis. He was discharged from the hospital March 23.

The patient was then lost track of by me and could not be traced, as he had moved from the residence he gave, until April 1, 1916, when he walked into my office. After leaving the hospital he had worked steadily as a pedler until recently, when the hemorrhages from the telangiectases on his face, tongue and nose prostrated him so severely that he found himself in a very much weakened condition and unable to work. A few days later I had a Wassermann and luetin test performed on him, with negative results in both instances. The internal administration of iron chlorid has yielded him some improvement.

Mrs. A. T., aged 33 years (Figs. 1 and 3, III, 24), first seen by me at my office on March 20, 1909, complained of bleeding from the face. The family history is given in the recital of the history of the previous case. The past history shows typhoid fever when the patient was 24 years old, of nine weeks' duration, with complete recovery. Two years previously she injured her thigh severely in an explosion. Her menses have been regular, occurring every twenty-eight days, with a duration varying from three to seven days and with slight pain. She has been married for ten years and has had three children, the age of the eldest being 8 and of the youngest 3. The labors have been instrumental and difficult.

Present Illness: She has bled from the nose ever since she can remember. The epistaxis begins and stops spontaneously. The intervals between the bleedings have varied from three to four weeks up to two or three months. One year ago ergot internally seemed to check them. At this time she first began to bleed from the tongue, pharynx and hard palate. These hemorrhages are also well controlled by ergot and the local application of tincture of ferric chlorid. Ten days ago the bleeding began to be more profuse and would come on especially during eating, when mastication would frequently remove a clot from the tongue.

Physical Examination: The patient was a well-nourished, well-built woman. The lips and mucous membranes were quite pale in color. The tongue was clean. On the dorsum of the tongue, near its middle aspect and slightly to the right, was a raised nodular angioma, split pea in size, with a flattened summit, from the center of which a pinpoint opening of a blood vessel was seen. The mucous membranes of the hard palate showed a number of telangiectases, pinhead in size. They were also seen about the vicinity of the cheek bones, one on the left side of the nose to the right of the bridge and 1.5 cm. from the tip being of the nodular type and split pea in size. The others varied from pinpoint to pinhead in size. All were bright red in color and did not blanch on pressure. On the right hand, near the styloid process, was seen a spot pinhead in size. The heart revealed a soft systolic murmur at the apex,

which is not transmitted outward and was heard with increasing intensity on passing upward, being loudest at the pulmonic area. There was a soft systolic murmur in the vessels of the neck.

Blood examination showed red blood corpuscles, 4,304,000; leukocytes, 7,750; hemoglobin (Dare), 55 per cent.; blood platelets (Pratt's method), 204,788; coagulation time (Biffi-Brook's method), two minutes. Differential count of 500 leukocytes gave polymorphonuclears, 61.8 per cent.; lymphocytes, 26 per cent.; large mononuclears and transitionals, 9 per cent.; eosinophils, 2 per cent., and mast cells, 0.4 per cent. There were no myeloblasts, no normoblasts or megaloblasts, no poikilocytosis, and but slight anisocytosis.

Subsequently the bleeding increased in severity and frequency so that the patient has occasionally been much prostrated by the secondary anemia thus induced. The use of rest, iron and arsenic has been of little avail.

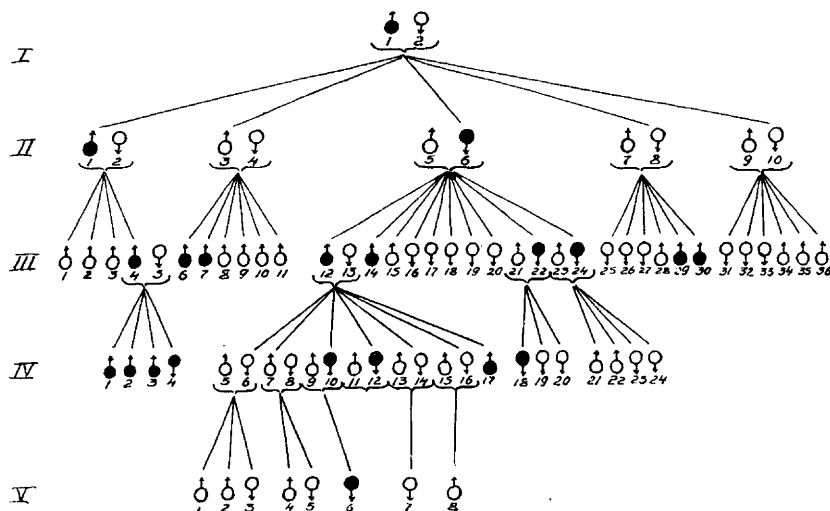


Fig. 3.—Family 26, Steiner 1. The heavily shaded characters in this and all succeeding figures indicate the members of the family affected, while the Roman numerals at the margin indicate the successive generations.

Mrs. D. E., aged 25 years (Figs. 2 and 3, IV, 12), was first seen by me at my office on April 26, 1909, complaining of bleeding from the nose. The family history is given in the recital of the history of the first case. The past history shows influenza every winter ever since she can remember. She had brain fever when very young and measles when she was a child. Her menses began at 14 and were regular every twenty-eight days until five or six years ago, since which time they have come a few days more frequently. She has much pain during the first two days.

Present Illness: Nine years ago she first began to bleed from the nose. The hemorrhages have come on since then five or six times a day, sometimes once a week or more rarely once a month, occasionally there is a two or three month interval. A little worry or excitement will generally bring on an attack, which frequently lasts five or ten minutes. She has never used anything to check the hemorrhages except cold water. Last summer she first began to bleed from the tongue and has bled thus about five times in all.

Physical Examination: The patient was a well-nourished, well-built woman. The lips and mucous membranes were of good color. Over the cheeks, chin, mucous membrane of the lower lip and the tongue were telangiectases varying

in size from a pinhead to split pea in size of which there were two on the tongue which were slightly raised. Some were of the spider type. All were cherry red in color. The heart was negative on examination. A Wassermann reaction on the blood and spinal fluid was later performed with negative results. Blood examination showed red blood corpuscles 4,000,000, white blood corpuscles 10,000, hemoglobin 75 per cent. The urine examination was negative. Subsequently the bleeding, especially from the nose, has increased in severity. The cauterization of the bleeding areas has been of little avail.

FAMILY 27 (Fig. 4) (Steiner 2).—Recurring epistaxis occurred in three members of the family for two generations; telangiectases with epistaxis were present in the only member of the family seen.

E. N., aged 41 years (Hospital No. 66927), was admitted to the Hartford Hospital on my medical service, March 17, 1913, complaining of being run down (Fig. 4, II, 8).

Family History: Her father had died of old age, her mother of pneumonia. Of her four sisters and one brother, one, a sister (II, 5), aged 47, had been subject to attacks of epistaxis all her life, but especially during childhood. She is married but has no children. She resides in Stockholm, Sweden. Another sister (II, 7) has a son (III, 7), aged 17, who has had frequent attacks of epistaxis since early childhood. There are no others in the family subject to epistaxis.

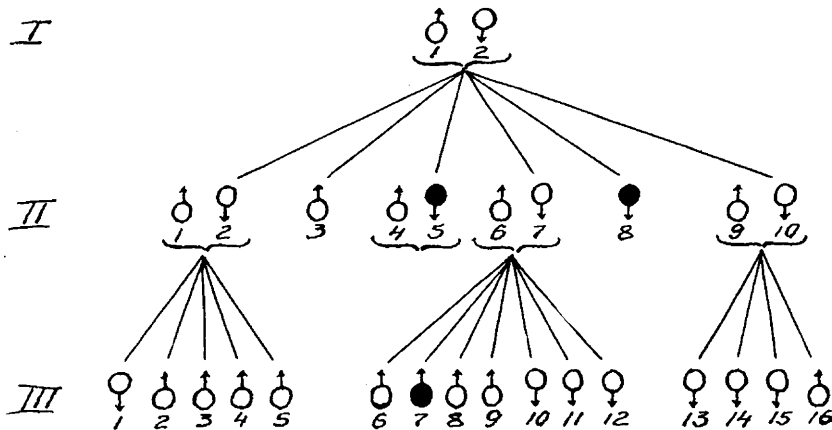


Fig. 4.—Family 27, Steiner 2.

Past History: The patient had measles at 8 years of age. Her menses began at 14, were regular every twenty-eight days, with a duration of six or seven days. The flow was always very profuse.

Present History: When she was about 25 years old, she began to have attacks of moderate epistaxis the day preceding the onset of each menstrual period. Her gums, however, have always bled easily when she cleans her teeth. Twelve years ago she emigrated to this country from Sweden, and two or three years later she began to have red spots on her cheeks. Two years later she noticed them on her hands and wrists. Occasionally she has had slight attacks of dizziness during the past few months. For the past week she has had one to three attacks of severe epistaxis daily, followed by sensations of weakness. The hemorrhages last from ten to fifteen minutes and cease spontaneously.

Physical Examination: The patient was a rather poorly nourished, slenderly built woman. The lips and mucous membranes were quite pale. Over the ears, cheeks, mucous membranes of the lips, tongue and cheeks, wrists,

hands and feet were telangiectases, mostly of the nodular type, pinhead in size, but some were of the spider variety. Some were also seen under the nails of the middle and index fingers and thumb of the left hand. The lungs were negative. The heart was not enlarged, but revealed a presystolic thrill and murmur at the apex, sharply localized. A systolic murmur was also here audible, which could be heard outward to the anterior axillary line and upward to the base, but in both directions with decreasing intensity. The pulmonic second was accentuated.

Blood examination showed red blood corpuscles 2,560,000; white blood corpuscles 5,200; hemoglobin (Dare) 55 per cent.; coagulation time four and a half minutes. Differential count showed polymorphonuclears, 69 per cent.; lymphocytes, 15 per cent.; large mononuclears and transitionals, 24 per cent., and eosinophils, 2 per cent. There were no myeloblasts and but one normoblast seen. There was marked anisocytosis and moderate poikilocytosis.

The patient refused to have the bleeding points in her nose cauterized. Her treatment consisted in the use of rest and iron. After a week's sojourn in the hospital, she left on March 24 to set sail for Sweden from New York on the following day.

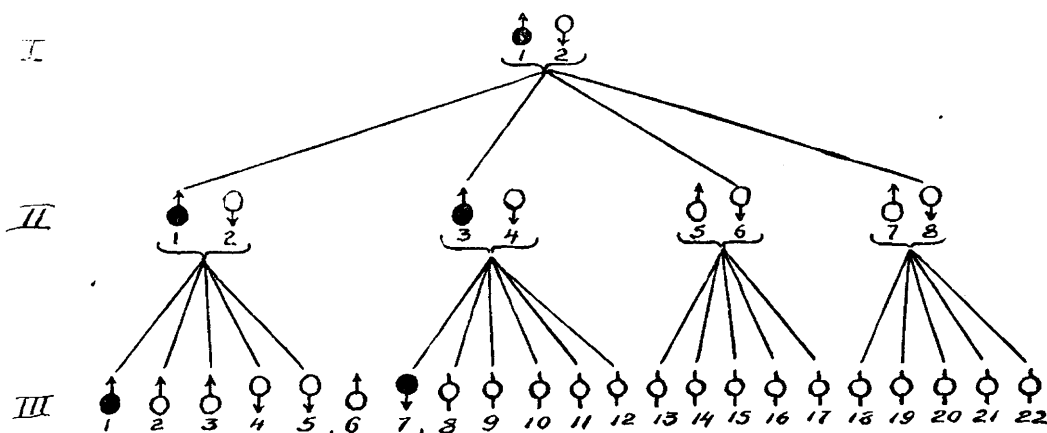


Fig. 5.—Family 28, Steiner 3. The character with the arrow stem both at the top and bottom of the circle in this family tree and in succeeding figures indicates that the sex is not stated.

FAMILY 28 (Fig. 5) (Steiner 3).—There was a history of epistaxis and telangiectases in the family for three generations.

C. L., aged 62 (Hospital No. 86348), was admitted to the Hartford Hospital on my medical service March 15, 1916, complaining of a cough (Fig. 5, II, 3).

Family History: His father (I, 1) who died at 92, had telangiectases and nasal hemorrhages, but further history about him was not obtainable. A brother (II, 1) had a similar condition, as well as his brother's son (III, 1). Of the patient's six children, a daughter (III, 7) suffers from occasional attacks of epistaxis.

Past History: The patient had always enjoyed good health. A few weeks before he entered the hospital, he had a slight operation on his tongue for what he says was a cancer. He had bled from the nose occasionally since boyhood, but recently the attacks had lessened in severity and frequency.

Present History: Four weeks ago he contracted a bronchitis and has coughed considerably ever since. With the cough he expectorates a little clear tenacious sputum. There are no other symptoms.

Physical Examination: The patient was a well-developed, well-nourished Italian. There was evidence of a slight operation on his tongue on the left side. The edges of the healed wound were not ulcerated but felt indurated. The teeth were in very bad condition and the breath was very foul. The lungs revealed medium, moist râles at both bases, posteriorly and in the left axilla. Over his forehead, cheeks and nose pinpoint, nodular and spider angiomas were noted. The largest one of these was located on the left cheek, 4 cm. below a line drawn from the middle of the lower eyelid and 1.5 cm. from the left ala nasi. Under the use of expectorants his cough lessened in frequency, became looser in character and finally left him. A Wassermann reaction was performed on his blood, withdrawn from a vein at his left elbow, with negative results. His teeth were cleaned by Dr. W. N. Butler, one of our dental surgeons, and the patient was discharged from the hospital in good condition on April 12, 1916.

FAMILY 1 (Fig. 6) (Legg, 1876).—There was a history of epistaxis in three generations; developmental telangiectases were noted on the patient's face and trunk, but there was no history of hemophilia.

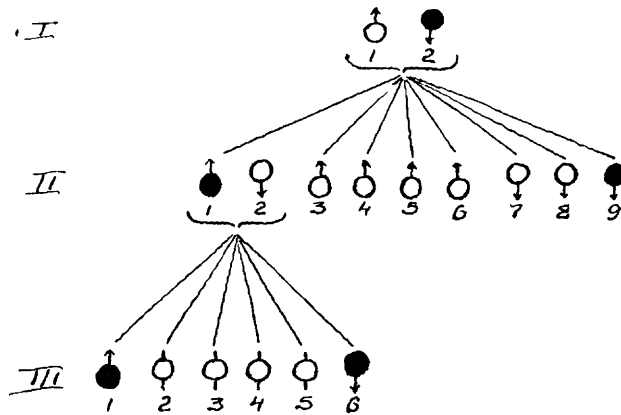


Fig. 6.—Family 1 (Legg).

The patient, II, 1 (Fig. 6), aged 65, gave a history of attacks of epistaxis since boyhood. He has also shown a tendency to bleed profusely from traumatic causes. His mother (I, 2) died from loss of blood and dropsy. One sister (II, 9) is subject to epistaxis. Of his six children a son, aged 27 (III, 1), and a daughter, aged 22 (III, 6), suffer from epistaxis. The patient has numerous small nevi over his face, forehead and various parts of his trunk which first appeared about his 41st year. Violent fits of anger as well as excessive drinking bring on attacks of epistaxis. When seen he had bled every day for six weeks.

FAMILY 2 (Fig. 7) (Chiari, 1887).—There was a history of epistaxis in four generations, with multiple telangiectases on the skin and mucous membranes, a condition diagnosed as hemophilia.

Two sisters (III, 5 and 7) stated that their grandmother (I, 3) and her two brothers (I, 1 and 4) suffered from childhood with frequent and severe attacks of epistaxis, yet lived to ripe old ages. Their mother (II, 2) also had the same difficulty and died at the age of 47 from dropsy, which apparently developed after a long period of nosebleed. Two brothers and one sister (III, 1, 2 and 3) were similarly affected with these severe attacks of epistaxis, but the sister had apparently recovered from these attacks, except at each menstrual period, when the epistaxis was insignificant. The two sisters mentioned above (III, 5 and 7) have bled since childhood from the nose, and one

of them once bled for a long time from the gums. Both of them have telangiectases on their faces and the upper part of their bodies. These are also seen on the mucous membranes of their noses, tongues and lips. III, 5 had four children, the youngest of whom, aged 4, bleeds from the nose. III, 7 had a son, aged 6, who has had epistaxis for four years.

FAMILY 3 (Fig. 8) (Chiari, 1887).—There was history of severe epistaxis in three generations, with multiple telangiectases on the skin and mucous membranes.

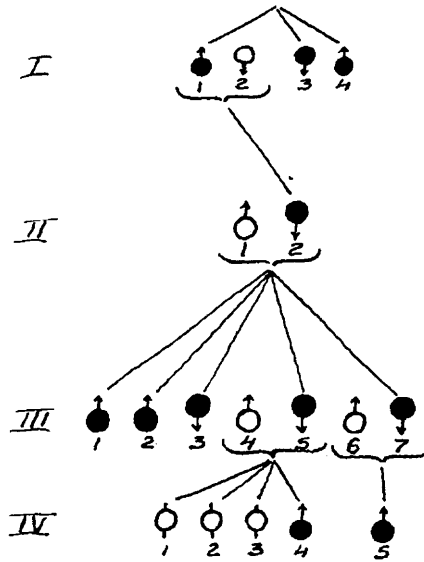


Fig. 7.—Family 2 (Chiari 1).

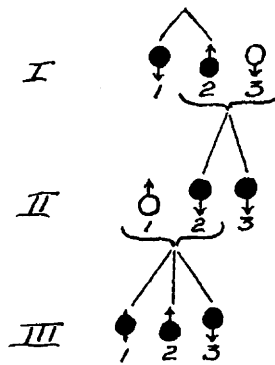


Fig. 8.—Family 3 (Chiari 2). The patient III, 1 died from severe epistaxis.

II, 3 had suffered from childhood with severe attacks of epistaxis. She was about 30 years old when first seen by Chiari and exhibited telangiectases on the skin and mucous membranes of the nose, tongue and lips. She stated that her father (I, 2) and her paternal aunt (I, 1) had had the same trouble. Her sister (II, 2) was likewise affected, as well as her sister's three children (III, 1, 2 and 3). One of them had died some years previously from a severe nosebleed.

FAMILY 4 (Fig. 9) (Rendu, 1896).—There was history of severe epistaxis in two generations; the condition was made a distinct clinical entity by Rendu.

II, 1, aged 52 years, had had severe daily recurring epistaxis for three weeks. His father (I, 1) had died of dysentery, with repeated crises of melena, at the age of 55, and his mother (II, 2) was subject to recurring epistaxis. One brother (II, 2) had also suffered from abundant and recurring epistaxis. Telangiectases were noted on the skin of the face and mucous membranes of the mouth and tongue of the patient. They were also seen, but in less numbers, over the neck and chest.

FAMILY 5 (Fig. 10) (Osler, 1901).—There were attacks of epistaxis from childhood, seven members of the family being subject to it; there were telangiectases on skin of face and mucous membranes of nose and mouth.

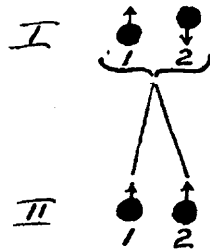


Fig. 9.—Family 4 (Rendu).

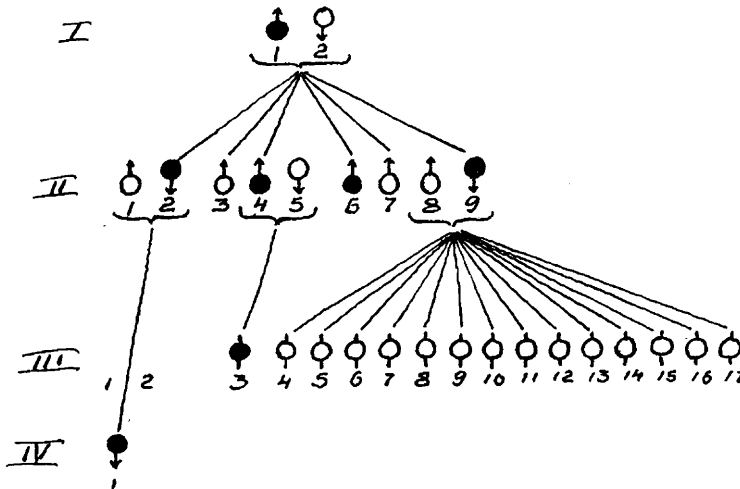


Fig. 10.—Family 5 (Osler 1).

II, 4 began to have attacks of epistaxis in his 10th year. They were not very severe, but recurred almost every day until he was 37 years old, when he was unable to work for nearly three years on account of the weakness and anemia induced by the bleeding. He was 49 years old when first seen by Osler, and exhibited telangiectases on the ears, nose, cheeks, lips and tongue. His father (I, 1) had had attacks of epistaxis, which were never dangerous but very frequent. His oldest sister (II, 2) had bled from childhood from the nose and her granddaughter (IV, 1) had had epistaxis frequently. Another sister (II, 9), also, had bled from the nose and mouth since childhood. His child (III, 3) had likewise occasionally bled from the nose, while a brother

(II, 6) gave a history of epistaxis from childhood with telangiectases of the skin and mucous membranes, which bled at times. This last patient finally died of cancer of the stomach, as revealed by necropsy.

FAMILY 6 (Fig. 11) (Osler, 1901).—Recurring epistaxis had occurred in the members of the family from the 10th year; there were multiple telangiectases of the skin and mucous membranes of the nose and mouth, and telangiectases but no epistaxis in the next generation.

Patient I, 1, aged 49 years, had had epistaxis from his 10th year, varying greatly in severity. From his 18th to his 21st year he was much better, but then the attacks came on again with increased severity. When a boy he first noted the telangiectases, which have increased during the past seven or eight years and are situated on his ears, forehead, cheeks, lips, back, chest, abdomen and hands. One of his sons showed vascular nevi (II, 1).

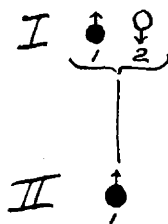


Fig. 11.—Family 6 (Osler 2).

FAMILY 7 (Fig. 12) (Josserand, 1902).—Telangiectases and epistaxis were observed in four members of the family during two generations.

Patient II, 1, aged 56 years, had suffered from epistaxis from childhood. Of late years she had had hemorrhages also from the lips, gums and tongue. Telangiectases were observed on her cheeks, lips, tongue and palate and less markedly on her neck, breast, back and arms. Her father (I, 1) and one brother (II, 2) had suffered from recurrent epistaxis from early childhood, while another brother (II, 3), in addition to this symptom, presented facial telangiectases.

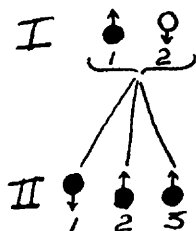


Fig. 12.—Family 7 (Josserand).

FAMILY 8 (Fig. 13) (Brown-Kelly, 1906).—There was severe recurring epistaxis with multiple telangiectases of the skin and mucous membranes in the family for three generations; death of one member occurred from syncope induced by a prolonged epistaxis.

The patient, II, 2, aged 41 years, had suffered from severe recurring epistaxis. Her father (I, 1) had died at 62 years of age from the effects of frequent attacks of epistaxis. A sister (II, 3) has suffered from frequent hemorrhages from her lips, mouth and nose, and has multiple telangiectases on the skin of her face and hands, and the mucous membranes of her nose and mouth. A few have lately developed on the scalp and occasionally give rise to

bleeding when the hair is combed. Her daughter (III, 1) bleeds from her nose and has red spots on her face. The patient (II, 2) finally died from an attack of syncope induced by severe and persistent epistaxis.

FAMILY 9 (Fig. 14) (Hawthorne, 1906).—There was epistaxis in three generations, with telangiectases, a marked family history of these symptoms.

II, 3, aged 49 years, had suffered from childhood with bleeding at the nose, which once required plugging of the nostrils. Her father (I, 1) and her sister (II, 1) had similar attacks and all had telangiectases on their faces.

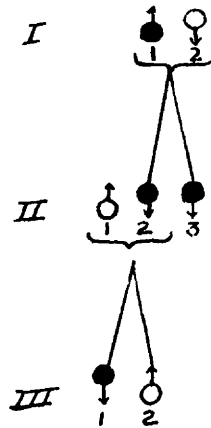


Fig. 13.—Family 8 (Brown and Kelly).

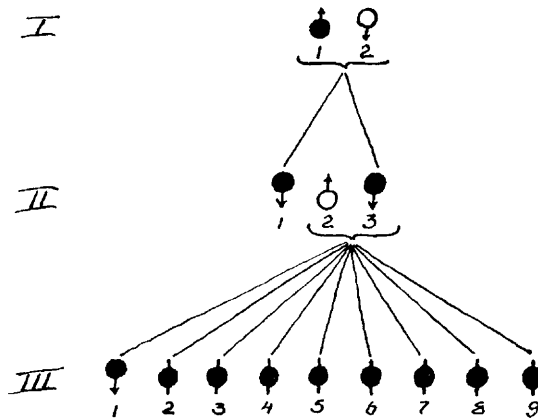


Fig. 14.—Family 9 (Hawthorne).

A few were also seen on the patient's fingers (II, 3) and all of her nine children were subject to recurring nose bleed.

FAMILY 10 (Fig. 15) (Osler, 1907).—Hemorrhages occurred from nose and mouth of one member of the family from the 10th year; there were multiple telangiectases on the skin of his face, ears and lips. A similar condition was observed in three generations.

The patient, III, 1, aged 53 years, is a physician, whose grandfather (I, 1), father (II, 1) and one sister (III, 3) had red spots on their faces. His son

(IV, 1) is subject to attacks of epistaxis which have been frequent but not profuse. III, 1 has bled with great profuseness from his nose and from the telangiectases on his head, face, ears, arm and mucous membranes of his mouth.

FAMILY 11 (Fig. 16) (Parkes-Weber, 1907).—This family gives a history of recurring epistaxis in four generations; telangiectases also were observed in four of the eight members affected with telangiectases.

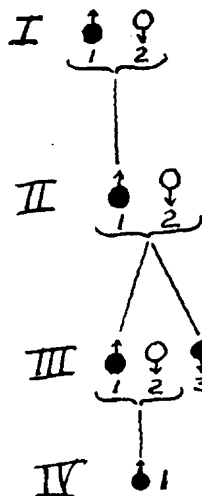


Fig. 15.—Family 10 (Osler 3).

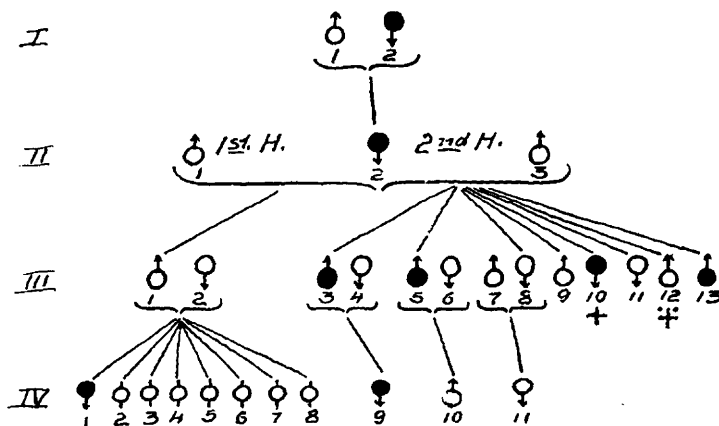


Fig. 16.—Family 11 (Parkes-Weber). The daughter, III, 10 suffered with epistaxis only. The son, III, 12, aged 3 years, died of "brain trouble."

The patient, II, 2, aged 60 years, first noted telangiectases on her face when she was 42 years old. Recurrent attacks of epistaxis had begun a few years earlier. During the last few years they have come on every two or three weeks and are more severe than formerly. The telangiectases are located on her face, ears, lips, tongue, mucous membranes of her mouth and nose and the conjunctival surface of the four eyelids. They are also observed on the

anterior surface of the epiglottis and on the fingers and under the finger nails. All are punctiform save one on the right cheek, which is of the spider nevus type. Her mother (I, 2) was subject to attacks of epistaxis and had one or two spots on her face. By her first marriage, she had a son, whose boy (IV, 1), aged 9, suffers from epistaxis. By her second marriage she has three sons (III, 3, 5 and 13) and one daughter (III, 10) who suffer likewise from epistaxis and two of these sons (III, 3 and 5) have multiple angiomas of the skin and mucous membranes. The oldest son (III, 3) is married and has a boy (IV, 9) who is subject to attacks of epistaxis.

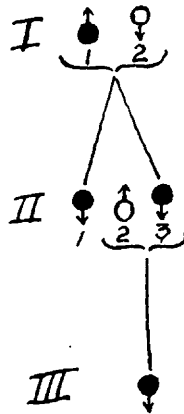


Fig. 17.—Family 12 (Phillips).

FAMILY 12 (Fig. 17) (Phillips, 1908).—Recurring hemorrhages from the nose and mouth occurred in this family in three generations; there were multiple telangiectases of the nose, tongue and buccal mucous membrane.

The patient, II, 3, aged 56 years, was subject to bleeding from childhood from the mouth and more recently from the nose. Her father (I, 1) suffers from violent epistaxis and bleeding from the tongue, while a sister (II, 1) died from hemorrhage of the gums. The patient (II, 3) has a daughter who has telangiectases on her tongue and recently has had attacks of epistaxis. The patient exhibits telangiectases on the mucous membranes of the tongue and nose, as well as on the buccal mucous membranes.

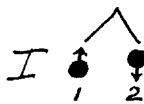


Fig. 18.—Family 13 (Waggett).

FAMILY 13 (Fig. 18) (Waggett, 1908).—There was severe epistaxis in brother and sister; multiple telangiectases of the skin of the face and the mucous membranes of the nose and mouth.

The patient, I, 1, aged 53 years, has had hemorrhage from the nose frequently since his 20th year. He has likewise had hemorrhages from telangiectases on his face and lips. These telangiectases are also seen on the mucous membranes of his tongue and nose. His sister (I, 2) has the same symptoms.

FAMILY 14 (Fig. 19) (Ballantyne, 1908).—This was a family of eight Hollanders who presented telangiectases in five of the six members examined.

The mother (I, 2) of a family of six children exhibited telangiectases on the conjunctiva of the right upper lid, the cheeks, the nose and the lips, but gave no history of hemorrhages from any of them. Her only daughter (II, 2) showed similar spots in almost similar locations, but has one also under the nail of the fourth finger of her right hand. She has bled from the nose rather frequently. A brother, II, 3, had no spots, but was subject to attacks of epi-

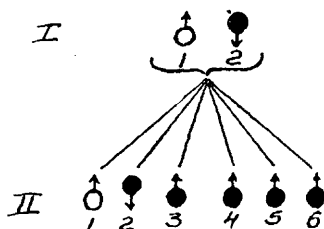


Fig. 19.—Family 14 (Ballantyne).

staxis. Another brother, II, 4, had a few telangiectases on the tip of his nose, inner surface of his lower lip and the tip of his tongue. The brothers, II, 5 and 6 showed small elevated red spots about the tip of the tongue as II, 4 did.

FAMILY 15 (Fig. 20) (Gottheil, 1908).—There were telangiectases in two generations; epistaxis in three.

The patient, II, 5, aged 40 years, has bled periodically and spontaneously from his nose, tongue and lips as long as he can remember. The hemorrhages

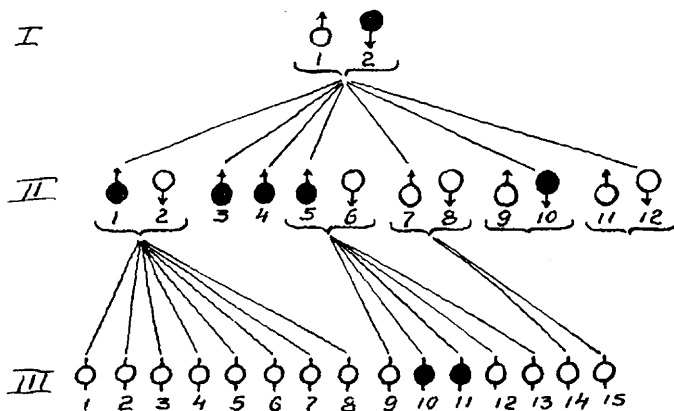


Fig. 20.—Family 15 (Gottheil).

vary in frequency from several in one day to one a week, but recently they have come on more often. His mother (I, 2) had spots on her lips and was said to have died from hemorrhages twenty-seven years ago. His brothers (II, 1, 3 and 4) have also had hemorrhages from the nose, as has also one sister (II, 10). None of them have had telangiectases. Two of his children (III, 10 and 11) have bled from the nose also.

FAMILY 16 (Fig. 21) (Hanes, 1908).—There were recurring hemorrhages in four generations of one family, and multiple telangiectases of skin and mucous membranes, but no symptoms of hemophilia.

The patient, II, 4, aged 53 years, had been subject to epistaxis from childhood. Generally her nose bleeds once or twice a week and sometimes every day. Since her 37th year the epistaxis has increased in severity, as well as the telangiectatic spots on her face, lips and tongue. Some are also seen on the ears, the hard palate and the gums of the lower jaw, as well as on the nasal mucous membranes. On the pads of the fingers and under the nails many small purple spots are visible. The conjunctiva of the right lower lid exhibits one of bright red hue. Her mother (I, 2), who died at 48 from heart disease with dropsy and cough, had suffered all her life with severe epistaxis. Her brothers (II, 1 and 2) were also troubled all their lives with a similar severe epistaxis and both showed red spots on their face and lips. One son (III, 1) has been greatly troubled from severe hemorrhages from telangiectases, which

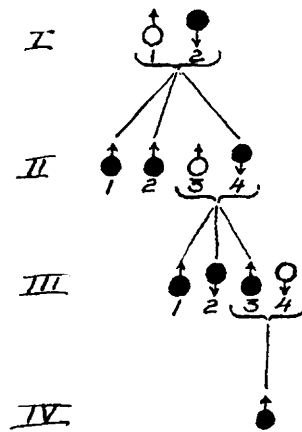


Fig. 21.—Family 16 (Hanes 1).

are numerous on his lips and tongue. Frequently he injures one of these labial or lingual telangiectases while eating, so that blood will spurt from the injured spot and render further progress with the meal impossible. Profuse hemorrhages from two telangiectases under the nail of the left middle finger have interfered greatly with his occupation as a fireman. Occasionally he has attacks of epistaxis. In every instance the hemorrhages result from slight traumatism. The other son (III, 3) had spots on his lips and face and had bled easily from his nose all his life. Her grandson (IV, 1), a son of III, 3, has two small red telangiectases at the mucocutaneous junction, but has never bled from the nose. Her daughter (III, 2), on the other hand, has frequently bled from the nose most profusely and presents on her face small pinpoint telangiectases. Several of a violaceous hue are seen on the lower lip and one bright red in color is noted on the dorsum of the tongue. The mucous membranes of the nose and the pads of the fingers and under the nails are other locations where these spots are found.

FAMILY 17 (Fig. 22) (Hanes, 1908).—There was epistaxis in four sisters and multiple telangiectases affecting chiefly the mucous membranes, but no symptoms of hemophilia.

The patient, II, 4, aged 46 years, had suffered extremely throughout childhood and youth from severe epistaxis, which frequently necessitated plugging of the nostrils. She also presented telangiectases under the eyes, at the bases of the alae nasi, on the conjunctivae, both lips, tip of the tongue, hard and soft palate, and nasal mucous membrane. She later developed an endocarditis. Her father (I, 1) had suffered from youth with recurring epistaxis, which came on daily during the later years of his life. One sister (II, 7), who

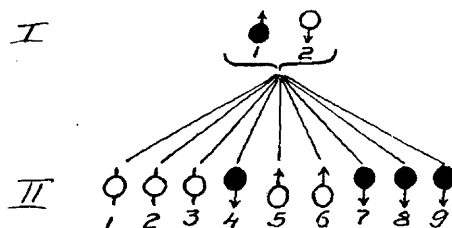


Fig. 22.—Family 17 (Hanes 2).

died at the age of 27, during childbirth, had bled profusely and frequently from the nose. Another sister (II, 8) had bled almost daily as far back as she could remember. She also presented definite telangiectases on her left cheek, lips, tip of tongue and nasal mucous membrane. Another sister (II, 9) bleeds two or three times weekly from the nose, but never very severely. She has a few telangiectases on the lips, tip of the tongue and nasal mucous membrane. Her two brothers (II, 5 and 6) are practically free from the disease, but bleed readily from the nose during a cold or after traumatism.

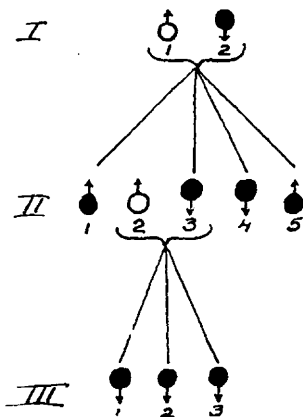


Fig. 23.—Family 18 (Laffont).

FAMILY 18 (Fig. 23) (Laffont, 1909).—A marked family history of telangiectasis, with a history of epistaxis in three out of the eight affected in the three generations.

The patient, II, 3, aged 48 years, noted eight years previously that telangiectases were appearing on the periphery of the scalp, the ear, the face, the breast and the back. Some had disappeared spontaneously. She had been subject since puberty to repeated attacks of epistaxis. Her mother (I, 2) had had, toward her 62d year, small telangiectases appear on her ear, neck, chest and

arm. There was no history of epistaxis in this instance. Her sister (II, 4) had similar spots on her ears, nape of neck and arm, but no epistaxis. Her elder brother (II, 1) had no telangiectases, but had spat up blood for about a year from varicosities of the pharynx, while her younger brother (II, 5) had some spots on his chest and back, but no hemorrhages. Her eldest daughter (III, 1) showed some telangiectases on her face and body, and had been subject to repeated epistaxis. Another daughter (III, 2) showed spots on the dorsal aspect of the forearm, the neck and trunk, while the third daughter (III, 3) presented spots on her face, neck, breast and back.

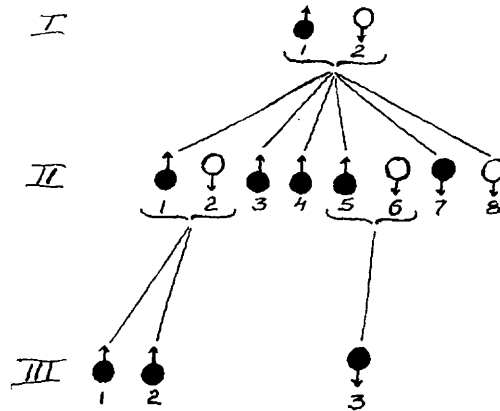


Fig. 24.—Family 19 (Langmead).

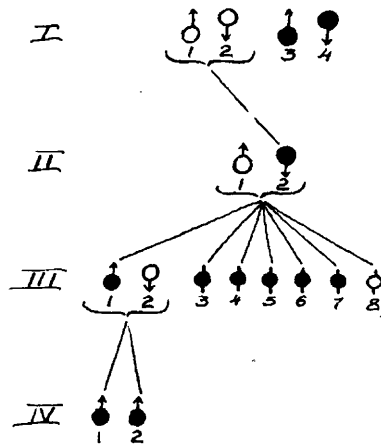


Fig. 25.—Family 20 (Audry).

FAMILY 19 (Fig. 24) (Langmead, 1909).—Telangiectases were noted in eight members of a family for three generations, epistaxis being observed in eleven instances.

The patient, II, 1, aged 68 years, was subject for about twenty years to frequent epistaxis. Occasionally telangiectases on his face and tongue have burst out bleeding. The telangiectases are seen on his face, lower lip, tip of tongue, palate and a few on the neck, chest and back. His father (I, 1) was

affected with epistaxis and telangiectasis, but his mother (I, 2) had epistaxis only. His three brothers (II, 3, 4 and 5) were subjects of epistaxis and telangiectasis, as was also one sister (II, 7). One son (III, 1) has nevoid patches and epistaxis, while the other (III, 2) has epistaxis only. A niece (III, 3) has similar patches and recurring epistaxis.

FAMILY 20 (Fig. 25) (Audry, 1911).—Epistaxis and telangiectases were noted in four generations.

The patient, III, 1, aged 70 years, had had attacks of epistaxis for many years, but lately they had increased greatly in frequency. Numerous telangiectases were seen on his face, lips, tongue and palate, and more sparsely on his trunk and upper extremity. His mother (II, 2) had epistaxis and telangiectases, as had also a great uncle and a great aunt. Five of the patient's brothers and sisters were similarly affected, as well as his two sons. Several nephews and nieces also had the same condition.

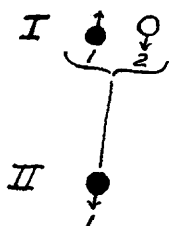


Fig. 26.—Family 21 (Osler 4).

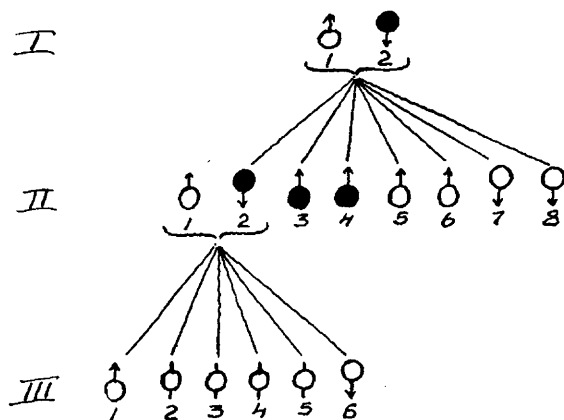


Fig. 27.—Family 22 (Van Wagenen).

FAMILY 21 (Fig. 26) (Osler, 1911).—Telangiectases were observed in two members of a family for two generations.

The patient, II, 1, aged 35 years, had suffered from childhood from recurring epistaxis. She also had shown later some telangiectases, but they had increased considerably during the last six years. They were situated on the nasal mucous membranes, the tongue and the lips. One was also seen under the nail of the left index finger. Her father (I, 1) had suffered for many years from severe recurring epistaxis and his face was covered with telangiectases.

FAMILY 22 (Fig. 27) (Van Wagenen, 1912).—Epistaxis and telangiectases were observed in four members of a family during three generations.

The patient, II, 2, aged 32 years, has had bleeding from the tongue and nose during the past nine years. Telangiectases were seen on the nasal mucous membranes, tongue, face and arms. Her mother (I, 2) has had frequent attacks of epistaxis since early childhood and two of her brothers (II, 3 and 4) have had a similar history.

FAMILY 23 (Fig. 28) (Sequeira, 1913).—Epistaxis and telangiectases were observed twice in two generations.

The patient, I, 2, aged 55 years, has noted red spots appearing on her face and fingers during the past five or six years. Occasionally she has had

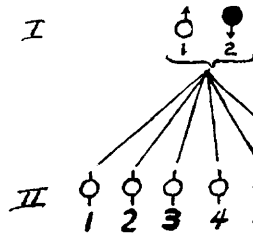


Fig. 28.—Family 23 (Sequeira).

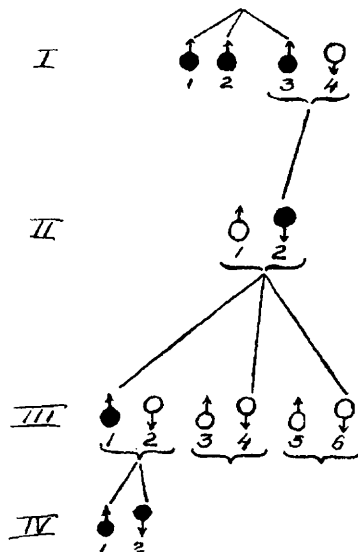


Fig. 29.—Family 24 (Gjessing).

hemorrhages from these spots. Telangiectases were also found on the tongue, mucous membrane of the lower lip, hard palate, uvula and the lower internal surface of the left labium majus. She had had also slight bleeding from the nose. One daughter (II, 6) gave a history of occasional attacks of epistaxis.

FAMILY 24 (Fig. 29) (Gjessing, 1915).—Epistaxis and telangiectases were observed for four generations, one member being affected also with endocarditis and hemorrhagic retinitis.

The patient, III, 1, 51 years of age, has had severe epistaxis from childhood, which lately has increased in frequency and violence. In his late twen-

ties telangiectases began to develop on his face and the mucous membranes of his mouth. They are now also seen on the ears, under the chin and on the neck, as well as on the nose, the tip and base of the tongue, the hard palate and the lower right eyelid. On the left arm two small ones are also noted, others under two finger nails and still others on the dorsal and volar aspects of the fingers. Besides the hemorrhages coming from the nose, bleeding has been observed from spots on his cheeks, tongue and eyelid. His maternal grandfather (I, 3) and the latter's two brothers (I, 1 and 2) had suffered from severe epistaxis, but it was not known whether they had facial angiomas. His two children (IV, 1 and 2) have suffered since childhood from severe epistaxis. The son (IV, 1) has telangiectases on his face, nasal mucous membranes, tongue, and hard palate. He is affected with a disease of the heart and has hemorrhagic retinitis. He has become so anemic from the hemorrhages that his hemoglobin has fallen to 25 (Tallqvist). The daughter (IV, 2) has a few small angiomas on her cheeks, which have appeared during the last two years.

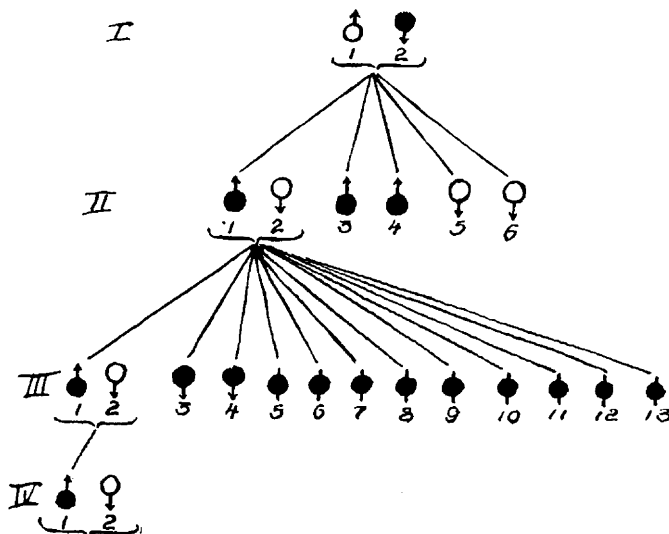


Fig. 30.—Family 25 (Hutchison and Oliver). Patients III, 5 to 13, inclusive, suffered with epistaxis only.

FAMILY 25 (Fig. 30) (Hutchison and Oliver, 1916).—There was a history of epistaxis in four generations, with telangiectases observed in three generations.

The patient, III, 1, aged 48, had had attacks of epistaxis for as long as he could remember, but during the last two years they had become more severe, especially in the morning. Telangiectases appeared first eight years ago and gradually increased in number. They were situated on the ears, cheeks, nose, lips, buccal mucous membranes, hard palate, tip and under surface of the tongue, the posterior pharyngeal wall, the soft palate, the uvula and the right hand. Spontaneous bleeding had occurred from those on the lips, and this was sometimes preceded by a feeling of soreness. He had also bled occasionally from the ears, about the fossa of the helix and from the corner of the right eye, as well as from the gums and the inside of the mouth. From the last situation the hemorrhages appeared especially after very hot drinks. About a year ago he bled from the rectum, on straining. His Wassermann reaction was negative. His father (II, 1) had had frequent spontaneous attacks of epistaxis from childhood and for the last six years has bled occasionally and

spontaneously from the spots on his nose. During the last five or six months he has bled somewhat from his gums. Two or three years ago he also had occasional bleeding from the rectum. For many years he has had telangiectases on his face. They were also noted on his ears, cheeks, sides of nose, lips, dorsum of tongue, hard palate and buccal mucous membranes, back and shoulders. His son (IV, 1) has had attacks of epistaxis as long as he could remember, but during the last two years they have become more infrequent. Some telangiectases were noted on his right cheek, the tip of his tongue, the mucous membranes of his lips and on his right shoulder. There was a definite history of nose bleeding in other members of the family (I, 2, II, 3 and 4, III, 3, 4, 5, 6, 7, 8, 9, 10, 11, 12 and 13).

CONCLUSIONS

From a consideration of the histories of these three families affected with hemorrhagic telangiectasis and those previously recorded we may conclude that heredity is the only important etiologic factor as yet discovered and that both sexes are equally affected and equally capable of transmitting the disease. Microscopically a developmental defect has been found in the dilated capillaries, as the elastic and muscle fibers appear to be wanting. The capillaries, consequently, are very liable to produce hemorrhages by their rupture, which is induced either spontaneously or by traumatism. The hemorrhages from the nose are generally seen early in life, while the telangiectases are a later development. The cautery in most instances offers the best results in the treatment of the bleeding telangiectases.