Arteriovenous Fistulas of the Lungs

By Samuel Baer, M.D., Albert Behrend, M.D., and Harold L. Goldburgh, M.D.

Two cases of pulmonary arteriovenous fistula (or aneurysm) are presented, one of which is reported in detail. Twenty-three other cases of pulmonary arteriovenous aneurysm or fistula previously reported in the American literature are reviewed, and the clinical, laboratory, radiologic, physiologic, pathologic and hereditary features of this condition discussed. Clubbing of the fingers and toes, polycythemia and cyanosis are almost constant findings. The importance of differentiating this condition from polycythemia vera and congenital heart disease is emphasized.

Published studies of pulmonary arteriovenous fistulas are not very common. There was no case of pulmonary arteriovenous aneurysm among the 111 cases of aneurysm of the pulmonary artery reviewed by Boyd and McGavack.1 Brief case reports of malignant pulmonary hemangiomas were published in 19312 and 1935,3 and in 1936 Bowers4 reported a fatal pulmonary hemorrhage due to a benign hemangioma in a newborn infant. It was not until 1938, however, that Rodes’s5 detailed description of a case called attention to its occurrence in adults. Since then the accumulated reports6–23 suggest that though pulmonary arteriovenous fistulas are relatively uncommon, the great majority have clinical and laboratory features which are typical and readily recognizable (table 1).

The disturbance is most apt to be found in the male, and all but five cases were recognized before the patient reached the age of 30 years. Cyanosis, clubbing of the digits, and polycythemia are the cardinal diagnostic findings. Dyspnea is an outstanding complaint and hemoptysis has occurred in one-third of the cases. Fainting, dizziness, convulsive seizures, and other cerebral manifestations are important secondary symptoms. At least half of the patients had telangiectasias or other vascular defects in the skin and mucous membranes. Abnormal pulmonary bruits were also present in 50 per cent of the cases.

As far as we are able to determine, the case we are reporting is the twenty-fourth on record in the American and English literature. Others, however, have been observed but remain unreported. We believe this disturbance is important not because of its rarity and bizarre features, but because many such cases are probably masquerading under a diagnosis of congenital heart disease or polycythemia vera. In view of the ever-present risk of severe or even fatal pulmonary hemorrhage, and the possibility of cerebral complications, the advantages of early diagnosis and surgical correction become apparent.

Case Report

J. H., a white man, age 21 years, was first seen at the age of 14, in January 1941. During routine examination for an upper respiratory infection, marked cyanosis of the lips, and cyanosis and clubbing of the fingers and toes were noted.

He had been quite normal at birth. No specific abnormality in infancy was recalled by his parents. Since 1940 they had noted considerable dullness of his lips and fingernails in cold weather and on vigorous exercise, especially swimming. Competition for the track team in high school had to be discontinued because of breathlessness. During a few months prior to the initial examination the patient had had a number of nosebleeds. The examination revealed a dusky cyanosis of the lips, fingernails, and toenails, with slight clubbing of the fingers and toes. The chest appeared completely normal. Neither by palpation nor percussion was the heart found to be enlarged; no thrills were felt. The heart sounds were of medium intensity and regular. A faint mitral systolic bruit was audible. Neither the liver nor spleen was palpable. The blood count revealed 107 per cent hemoglobin (Sahli) with 6,250,000 erythrocytes per cubic millimeter. The leucocyte and differential counts gave normal values. A diagnosis of polycythemia vera was made, and treatment consisting of weekly venesections was begun, 250 cc. of blood being withdrawn during each treatment.

From the Medical and Surgical Wards of Jewish Hospital, Philadelphia, Pa.

* Since this paper was submitted for publication, an excellent discussion of this topic, listing 44 cases in the American and foreign literature, has been published by Yater and others (J.A.M.A., No. 141, page 581, October 1949).
<table>
<thead>
<tr>
<th>Authors</th>
<th>Age</th>
<th>Sex</th>
<th>Cyanosis</th>
<th>Clubbing</th>
<th>Polycythemia</th>
<th>Dyspnea</th>
<th>Hemoptysis</th>
<th>Cardiac Enlargement</th>
<th>Brad</th>
<th>Oxygen Saturation</th>
<th>Other Symptoms</th>
<th>Other Telangiectasia</th>
<th>Location by X-Ray Examination</th>
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<td>74%</td>
<td>Moyer and Ackermann</td>
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<td>Pneumonecctomy</td>
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<td>0</td>
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<td>0</td>
<td>0</td>
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<td>0</td>
<td>0</td>
<td>0</td>
<td>Dizziness; blurred vision</td>
<td>Lips, face, nose</td>
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<tr>
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<td>M</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>Epistaxis; convulsions</td>
<td>Umbilicus</td>
<td>Bilateral</td>
<td>Lobectomies</td>
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</table>
At intervals during the next six years the patient underwent a series of venesections when his red blood count became too high, or the cyanosis too marked. At times the erythrocytes exceeded 8,000,000, despite reduction of the hemoglobin to 13 to 14 grams and the red cells to 4,700,000, the cyanosis was still present. The significance of this was not appreciated at that time.

In May 1947, detailed hematologic studies were made, and the diagnosis of polycythemia vera seriously questioned. In October 1947 the patient had a profuse hemoptysis and x-ray study of the chest was advised. On photofluorography a diagnosis of ad-
FIG. 3.—Simultaneous stethograms and carotid tracings made preoperatively. Lower curves in each record are carotid tracings. A, Stethographic record illustrating the late systolic murmur in the left axilla. B, Stethographic record taken in the right posterior thorax opposite the seventh dorsal spine, during inspiration. C, Tracing taken over the right posterior thorax, during expiration. Note the increase in intensity in the murmur.

Advanced tuberculosis of both lower lobes was made. Sputum examinations showed no evidence of tuberculosis. On fluoroscopy, peculiar nodular opacities were noted in the right lower lobe and at the left border of the heart. A roentgen study of the chest was then made (figs. 1 and 2). The bizarre nodular opacities previously noted were seen. The radiologist concluded that "in view of the patient's history
of polycythemia, the findings in the chest can be explained on the basis of a polycythemia vera. Large nodular shadows in the lungs, as well as pulmonary vascular dilatation, do occur in this condition. We believe, however, that a diagnosis of multiple pul-

The patient was admitted to Jewish Hospital on the medical service of Dr. H. L. Goldburgh, on November 18, 1947. On admission, the cyanosis of the lips, fingers, and toenails was noted. There was obvious clubbing of the fingers and toes. By palpa-

![Graph](https://via.placeholder.com/150)

**FIG. 4.—Electrokymograms and carotid tracings made preoperatively.** Lower curves in each record are carotid tracings. A, Electrokymograms taken over the middle left border of the heart and the lesion in the left lung (postero-anterior projection). The upper curve on the left is that of the middle border of the left ventricle. The upper curve on the right, which is quite similar, is that of the lesion in the left lung. B, Electrokymograms taken over the pulmonary artery (upper left) and the lesion in the right lower lung (upper right). The lesion in the lung showed an active movement of its borders corresponding to the arterial type of curve such as usually obtained over the pulmonary artery.

**TABLE 3.—Arterial Oxygen and Cardiac Output Studies**

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<td>9.7</td>
<td>9.0</td>
<td>12.9</td>
<td>14.7</td>
<td>13.1</td>
<td>11.2</td>
<td>12.5</td>
<td>15.6</td>
<td>15.8</td>
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<td>15.2</td>
<td>17.0</td>
<td>15.7</td>
<td>23.3</td>
<td>20.8</td>
<td>16.2</td>
<td>22.0</td>
<td>19.7</td>
<td>19.4</td>
<td>19.1</td>
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<td>Oxygen saturation (%)</td>
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<td>63.8</td>
<td>53.0</td>
<td>82.0</td>
<td>62.9</td>
<td>62.9</td>
<td>69.0</td>
<td>57.0</td>
<td>78.9</td>
<td>81.7</td>
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<tr>
<td>Hematocrit</td>
<td>about 45%</td>
<td>68.8%</td>
<td>66.6%</td>
<td>48.5%</td>
<td>55.5%</td>
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<tr>
<td>Cardiac output (liters/minute)</td>
<td>12.3</td>
<td>10.1</td>
<td>9.0</td>
<td>15.0</td>
<td>11.1</td>
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<td>Cardiac index (liters/minute/ sq.m.)</td>
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<td>7.0</td>
<td>6.0</td>
<td>5.7</td>
<td>9.5</td>
<td>6.7</td>
<td></td>
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</table>

Right lower lobectomy on 1-6-48; left lower lobectomy on 5-12-48; left lingulectomy on 9-28-48.

Determinations made in the Department of Physiology, Temple University Medical School, except those of 12-23-48 and 1-12-49 which were made in the Pulmonary Function Section, Graduate School of Medicine, University of Pennsylvania.

* After inhalation of 100 per cent oxygen.

monary cavernous angioma or arteriovenous fistula is more likely. Secondary polycythemia is part of the clinical syndrome of this disease. Angiocardiography could be used to prove the diagnosis. With this diagnosis in mind hospitalization was advised for confirmation and possible surgical therapy.
liver and spleen were not felt. The presence of moderate cardiac enlargement was demonstrated by both orthodiagram and x-ray study of the chest, as were the shadows and nodular opacities previously seen in the lower lung fields. Some of the laboratory findings at that time are tabulated in table 2. We again noted the presence of deep cyanosis despite a relatively normal blood count. Arm-to-lung and arm-to-tongue circulation times were normal. Attempts to delineate the pulmonary shadows by intravenous Diodrast were twice unsuccessful.

Synchronous sound and carotid tracings and roentgenkymograms gave further evidence that these pulmonary lesions were due to vascular abnormalities (figs. 3 and 4). Arterial oxygen determinations and cardiac output by the ballistocardiographic method were obtained (table 3, and fig. 5).

The various procedures performed were considered confirmatory of the diagnosis of bilateral arteriovenous fistulas of the lungs. The patient was discharged on December 4, 1947, with the understanding that surgery would be undertaken early in January 1948. Two weeks after leaving the hospital he had a number of severe hemoptyses, and he was readmitted December 29, 1947.

After the pulmonary bleeding subsided, lobectomy of the right lower lobe was performed on January 6, 1948. (Details of the surgical problems encountered in this and later operations are being reported elsewhere.) At operation numerous dusky, pulsating vascular tumors were seen throughout the right lower lobe. The thrill felt on palpation disappeared upon compression or clamping of the pulmonary artery supplying the lower lobe. During the operation several large blood vessels were noted entering the inferior portion of the right lower lobe; these seemed to have penetrated the diaphragm.

Dissection of the lung specimen revealed marked dilation of the pulmonary artery and vein which had numerous large branches. These communicated

Fig. 5.—Ballistocardiogram taken on December 15, 1947 (30-gram weight calibration, Nickerson technic). Cardiac index averaged 7.0 liters per square meter per minute.

Fig. 6.—Roentgen-ray films taken in September 1948. Note the large A-V fistula in the lingual portion of the left upper lobe.
with a number of cystlike cavities 0.5 to 2.0 cm. in diameter. Each cyst communicated with two vessels, probably an artery and a vein. One could not always be certain of this, however, for even at the hilum there were so many vessels that veins and arteries could not be distinguished.

The patient did quite well postoperatively. Improvement was slow but continuous. Effort tolerance increased and the cyanosis became less, though it never completely disappeared. Repetition of the ballistocardiographic study showed a decrease in cardiac index to 6.0 liters per minutes. The preoperative oxygen saturation of 53 per cent rose to 82 per cent postoperatively, as seen in table 3.

![Fig. 7.—Roentgenograms of the chest taken postoperatively in November 1948.](image)

The clinical condition having improved sufficiently, lobectomy of the left lower lobe was contemplated for the early part of May. On April 22 however, the patient suddenly had a convulsive seizure and lapsed into unconsciousness. He was aphasic upon admission to the hospital, and had a right sided hemiplegia. The Babinski sign was positive on the right. His condition appeared critical. The temperature rapidly rose to 103 F. and a blood count revealed 7,500,000 erythrocytes and 26,000 leukocytes per cubic millimeter, and 16.7 grams of hemoglobin per 100 cc. of blood. Following a phlebotomy of 500 cc. and left stellate ganglion block with 1 percent procaine hydrochloride, there was a marked improvement. The speech returned within forty-eight hours and the right-sided paralysis rapidly disappeared. Convalescence continued satisfactorily, and on May 12, 1948, lobectomy of the left lower lobe was performed. Study of the left lower lobe revealed a number of arteriovenous communications, varying in size and similar to those found in the right lower lobe. The postoperative course was uneventful; the patient was discharged in twelve days. However, it was observed during the next few weeks that the cyanosis had not disappeared, that the oxygen saturation had decreased to 62 per cent, and that the red cells rose to over 6,000,000 per cubic millimeter. A persistent tachycardia of 110 developed and deep cyanosis continued. It was felt that we were dealing with some postoperative pulmonary complication, or possibly recurrence of the pulmonary arteriovenous communication. Roentgen-ray study in September revealed a large, circular, sharply demarcated shadow in the lower portion of the left upper lobe, overlying the left cardiac border (fig. 6). We then decided that we were faced with an expanded or new arteriovenous communication, which seemed much larger than any of those previously encountered. With a good deal of trepidation it was decided to attempt removal of this aneurysm. Anterior thoracotomy on September 28, 1948, revealed a large arteriovenous fistula involving the entire lingual portion of the left upper lobe, which was resected. Dissection of the specimen indicated it was larger than any individual fistula found in either of the lower lobes. The patient was discharged in relatively good condition on October 8, 1948 (fig. 7).
DISCUSSION

Before considering the clinical features of this condition, it might not be amiss to discuss some of the congenital aspects of pulmonary arteriovenous fistulas. Most observers are in agreement with Wodehouse who considered hemangiomas of the lung as "local manifestations of a generalized congenital tendency to maldevelopment of the blood vessels." It is more than just chance that in 50 per cent of the reported cases other vascular abnormalities, such as telangiectasias, capillary hemangiomas, and spider nevi, were exhibited. Rigid search will probably disclose other associated vascular anomalies in a high percentage of discovered cases. Moyer and Ackerman have recently reported in detail two cases of familial telangiectasias with pulmonary involvement. They reviewed the question of hereditary familial telangiectasias, and pointed out the relationship of pulmonary hemangiomas to the generalized vascular disease. It is also true, however, that an occasional patient is seen in whom the pulmonary abnormality is the only vascular disturbance present.

The familial incidence of this disorder is worthy of comment. Goldman in reporting one patient found a similar involvement in a brother, and bleeding tendencies in other members of the family. We have already referred to the report by Moyer and Ackerman of 2 cases in brothers. In the discussion that followed Maier and collaborators presentation a number of discussers, in commenting on unreported cases they had seen, emphasized the familial aspects of the condition. While preparing the report on our patient, we noted that his father had a number of telangiectasias about the lips. Roentgen-ray study revealed nodular and cylindrical shadows in the lung fields that were quite suggestive of pulmonary hemangiomas.

Clinical Picture. The published reports of this disturbance emphasize the occurrence of the diagnostic triad—cyanosis, clubbing of the fingers and toes, and polycythemia. Cyanosis is usually the first finding, and was present in 21 of the 24 cases. Clubbing and polycythemia were found in 20 cases. All the manifestations of this clinical syndrome are physiologic responses to the anoxemia. A variable quantity of venous blood is shunted from pulmonary artery to vein without going through the pulmonary alveoli. As a result there is decreased oxygenation of the arterial blood, and a chain of compensatory mechanisms takes place. There is an increase in blood volume, as in all other arteriovenous fistulas. Burchell and Clagett found it above normal in all of the 5 patients whose blood volume had been measured. Increase in hemoglobin concentration occurs and finally a rise in the number of circulating erythrocytes. The degree of oxygen unsaturation determines the compensatory hematologic mechanisms that are called into play. As the oxygen unsaturation and the polycythemia increase, cyanosis becomes more marked. Clubbing of the fingers and toes is undoubtedly a response to the oxygen unsaturation. Hemoptysis and cough occur as a result of oozing from or rupture of one of the aneurysms. It might be advisable at this point to emphasize the frequent occurrence of cerebral symptoms. Headache, dizziness, tinnitus, convulsive seizures, and hemiplegia have been reported in 50 per cent of the cases. The cerebral involvement may be extremely disturbing, as in our case. It is precisely these symptoms, together with the polycythemia, the epistaxis, the bleeding gums, and the corneal injection, that have led to the continued diagnosis of polycythemia vera. It seems logical to suggest that the oxygen unsaturation or the secondary polycythemia or both may be responsible for these cerebral manifestations, if we can exclude the possibility of intracranial vascular anomalies.

In addition to the cyanosis and clubbing of the fingers and toes, physical examination usually reveals pulmonary and cardiac murmurs. It is just this cyanosis, clubbing, and cardiac murmurs that so often leads to a diagnosis of congenital heart disease. Many observers have reported the presence of pulmonary bruits over the arteriovenous communications, as were found in our patient. Some have reported the murmur loudest at the height of inspiration, and others at expiration. The murmurs and the thrill at operation disappeared with ligation of the abnormal vessels.
There is considerable disagreement concerning the cardiac abnormalities present. Kennedy, Burwell, and Sidney have reported that peripheral arteriovenous fistulas produce an increase in cardiac work. If this continues long enough, cardiac hypertrophy and congestive failure develop. Most of the reports, however, suggest there is no cardiac disturbance present in these cases. Jones and Thompson stated that "the lack of any cardiac hypertrophy or pathology is due to the fact that the arteriovenous fistula is confined to the pulmonary circuit." Smith and Horton, Burchell and Claggett, Bisgard, and Moyer and Ackerman all have subscribed to this view. Moyer and Ackerman attributed the absence of cardiac enlargement (that usually accompanies peripheral arteriovenous fistulas) to low pressure in the pulmonary circuit. Maier and associates stated that the cardiac output is normal.

We are not completely in accord with these views. The cardiac output in our patient was above normal, as was the frontal cardiac area. Wodehouse has also reported a patient with cardiac enlargement. In our patient, there was a loud mitral systolic bruit that has decreased considerably since the last operation. The frontal cardiac area preoperatively (as determined by orthodiagram by two separate observers) was 30 per cent above the predicted normal. There has been decrease in the heart size since removal of the fistulas, so that the transverse diameter and frontal area are now within the predicted normal range. It is interesting to note that Burell and Claggett’s patient also exhibited decrease in heart size following removal of the abnormal vascular communication. It is our feeling that the degree of cardiac hypertrophy depends on the degree of arteriovenous shunt, the increase in blood volume, and the amount of increase in cardiac work that results. Eventually, in all cases, some increase in heart size should be manifest, and a subsequent decrease following surgical extirpation of the fistulas.

**Roentgen-Ray Findings.** The radiologic findings in these cases are characteristic, and constantly present. They consist of one or more circular, cylindrical, or nodular lesions occurring in the lung fields. It will be recalled that Hirsch originally reported nodular lesions in the lung fields occurring in polycythemia vera. It is quite possible that some of these shadows are due to arteriovenous fistulas, as suggested by Rodes.

These pulmonary lesions are chronic and usually nonprogressive. They may pulsate, or there may be increased hilar pulsations on the side of the lesion. The determination of the number of fistulas present is important in any consideration of surgical treatment. It must be realized that the shadows demonstrated radiologically are usually the largest fistulas, but not necessarily the only ones. We have observed, as did Moyer and Ackerman, that fairly large subpleural lesions may escape detection. In addition, small unsuspected lesions in one lobe may expand after removal of the larger fistulas in another lobe.

Special roentgen-ray technics have been used to delineate these pulmonary shadows more accurately. On planigraphic examination it is occasionally possible to trace some of the larger vessels as they arise from the hilum. Angiocardiographic studies have been successfully performed in a few cases, though our results with this procedure were not satisfactory. Good roentgenologic demonstrations of the lesions outlined with Diodrast have been published in conjunction with the case reports of Watson and of Moyer and Ackerman. Roentgenkymography may also help differentiate these lesions from nonvascular shadows, as was done in our patient (see fig. 4).

**Pathologic Findings.** A number of the published reports have contained excellent discussions of the pathologic findings in the specimens obtained at operation or necropsy. We need not dwell on this phase of the problem at any length. It seems relatively academic whether these lesions are considered aneurysms or fistulas or hemangiomas. The degree of arteriovenous shunt determines the clinical abnormalities that occur. In some of the dissected specimens it has been possible to demonstrate the course and nature of the abnormal communications. In many, however, the number and degree of the anastomoses have been so great that satisfactory pathologic evaluation has been impossible.
Prognosis. It is generally agreed that serious or fatal hemoptysis will ensue if these pulmonary lesions are not treated surgically. Four patients have died from pulmonary hemorrhage. All those that have been operated upon have done well postoperatively, and maintained improvement as long as they have been followed after their surgery. An occasional patient, however, has refused surgery and apparently gotten along well. Some of the reported cases have been symptomatically negative and been discovered accidentally. The father of our patient, for example, is now 52 years of age, and has never consulted a physician for a cardiac or pulmonary complaint. It seems logical to assume that the number and degree of arteriovenous communications will determine the symptomatology and the resultant treatment.

We have already referred to the cerebral and other associated complications of this condition. In one of Wodehouse's cases22 a brain abscess occurred from which Haemophilus influenza was cultured. Death followed operation for the brain abscess. Maier and his associates23 reported a case in which there was superimposed bacterial endocarditis. The patient responded to antibiotic therapy and was then successfully treated by lobectomy.

How well our patient will do is problematic. Since his discharge he has continued to gain weight and strength. His effort tolerance has improved, dyspnea is less, and cyanosis considerably decreased. The heart size has returned to normal and there is no polycythemia. The last x-ray examination of the chest (fig. 7) gave evidence that all the arteriovenous communications had been removed. The oxygen determinations, however, cast some doubt on this. The most recent arterial oxygen saturation still was only 81.7 per cent (table 3). Following inhalation of 100 per cent oxygen the saturation rose to 92.8 per cent, considerably less than the 99 per cent to 100 per cent saturation that should occur normally. According to Comroe22 and others, this type of response is characteristic of an arteriovenous shunt. Continued observation will be necessary to determine the location of any remaining arteriovenous communications. It is possible that previously undetected fistulas in the upper lobes may eventually expand and become troublesome, with the changed pulmonary hemodynamics.*

Summary

1. Twenty-three cases of pulmonary arteriovenous fistulas have been reviewed, and an additional case in a man of 21 years reported in detail.
2. Some of the physiologic observations obtained have been presented.
3. The congenital and familial aspects of the condition are emphasized.
4. The clinical features of arteriovenous fistula have been discussed, as well as the resemblance to and differentiation from polycythemia vera and congenital heart disease.
5. We believe that this condition is not nearly as rare as considered, and that its clinical features should render its recognition easy.

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* Since this paper was submitted for publication, the patient has developed moderate polycythemia again, and cyanosis. Re-examination has revealed a new fistula in the remaining right lung.
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Arteriovenous Fistulas of the Lungs
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