A Surgical Approach to the Problem of Chronic Pulmonary Artery Obstruction Due to Thrombosis or Stenosis *

ELLIOTT S. HURWITT, M.D., CLARENCE J. SCHEIN, M.D., HAROLD RIFKIN, M.D., ALVIN LEBENDIGER, M.D.

From the Surgical, Medical, and Laboratory Divisions, the Montefiore Hospital, New York City

SUDDEN unexpected death due to massive pulmonary embolization, with or without premonitory evidence of peripheral venous thrombosis, is a situation with which surgeons are all too familiar. In the face of an overwhelming episode which has not been immediately fatal, a few Trendelenburg operations have been attempted, with rare survival. There is, however, a separate group of cases of chronic obstruction of the pulmonary artery in which this mechanical block to emptying of the right side of the heart is the only significant intrathoracic abnormality, and in which the possibility of operative salvage has received little attention by surgeons. It is the purpose of this paper to present two such cases of chronic thrombotic occlusion of the pulmonary artery, in one of which a surgical attempt to relieve the obstruction was made. The rationale for and technical details of a planned surgical approach to this problem will be discussed. The concept will be elaborated that recognition and careful selection of these cases of subacute and chronic cor pulmonale, as well as of instances of stenosis or stricture of the pulmonary arteries, should result in broadening the present scope of cardiovascular surgery.

* Submitted for publication May 15, 1957.

The experimental work was done in the Henry and Lucy Moses Laboratory, and was aided by a grant from the New York Heart Association.

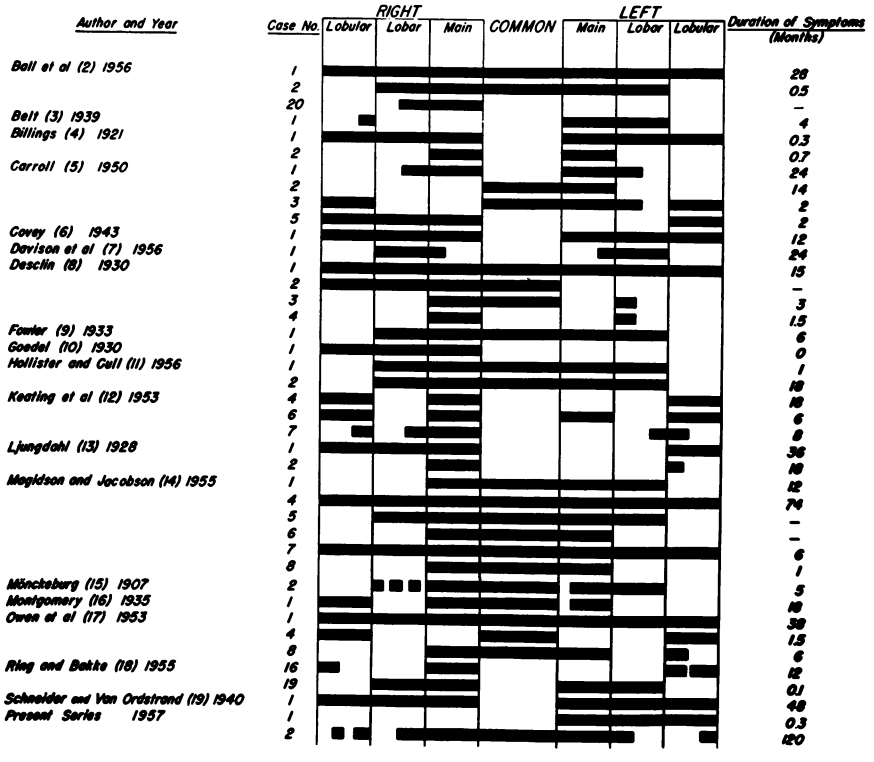
Chronic Thrombosis of the Pulmonary Artery

Approximately 300 cases of chronic thrombosis of the pulmonary artery have been reported. A recent review of 299 published cases disclosed the fact that 122 of these were secondary to congenital or acquired heart disease, pulmonary tuberculosis or other types of acute or chronic pulmonary or mediastinal disease, a blood dyscrasia, or an agonal phenomenon with overwhelming sepsis or severe debilitation.[1] There were 177 cases in which no concomitant cardiac, pulmonary, or mediastinal pathology was present, and in which the entire syndrome of progressive dyspnea, cor pulmonale, right heart enlargement, and right sided heart failure was due to the thrombotic occlusion within the pulmonary arterial tree. Since the cardiac manifestations in such cases are secondary to the pulmonary artery thrombosis, one may characterize this lesion as primary pulmonary artery thrombosis, despite the fact that the occlusion is most often a sequel to peripheral embolic thrombophlebitis or phlebothrombosis. Cases have also occurred as a result of severe trauma to the chest, and thrombosis on atheromata has been reported. Localized thrombosis has also been described in association with pulmonary arteritis and pulmonary artery aneurysms.

The documentation of the pathology in most of the 177 published cases compatible

TABLE I

LOCATION of THROMBUS in PULMONARY ARTERIAL TREE



with this concept of primary pulmonary artery thrombosis was insufficient to determine whether or not the extent and anatomic distribution of the thrombotic material in the pulmonary arterial tree was such as to permit surgical salvage by thrombectomy. Twenty-four of the cases were definitely inoperable, exhibiting multiple peripheral pulmonary emboli or thrombi, without involvement of the main trunk of the pulmonary artery or its major branches. Table 1 depicts the location of the thrombi in the 39 reported cases that included this information, plus the two cases in the present report (2–19). It is apparent that in approximately one-half of the cases the thrombus was either entirely central in location, or the central thrombus represented the major obstruction to the pulmonary outflow tract. Given the combination of an accessible site of thrombosis, and a substantial time interval between the onset of symptoms and death (Table 1), the potential for surgical salvage would seem to depend on an awareness of the problem on the part of all members of the medical profession.

Case 1. M. B., Montefiore Hospital admission number 77555, was a 62-year-old white female who underwent subtotal thyroidectomy at this hospital on December 29, 1955, for an adenomatous goiter (C. J. S.). She was discharged from the hospital on January 8, 1956, in good condition, and readmitted on January 11, 1956, 3 days later, with a 10-hour history of dyspnea, progressively increasing in severity. During the ensuing 11 days the clinical picture was dominated by acute distress, dyspnea, orthopnea, restlessness, and confusion. The neck veins were not distended, the lungs were clear, no abnormal cardiac signs were present, the liver was not enlarged, and there was no peripheral edema. The venous pressure was 150 mm. of water, rising to 200 mm. on right upper quadrant pressure. The circulation time, with so-

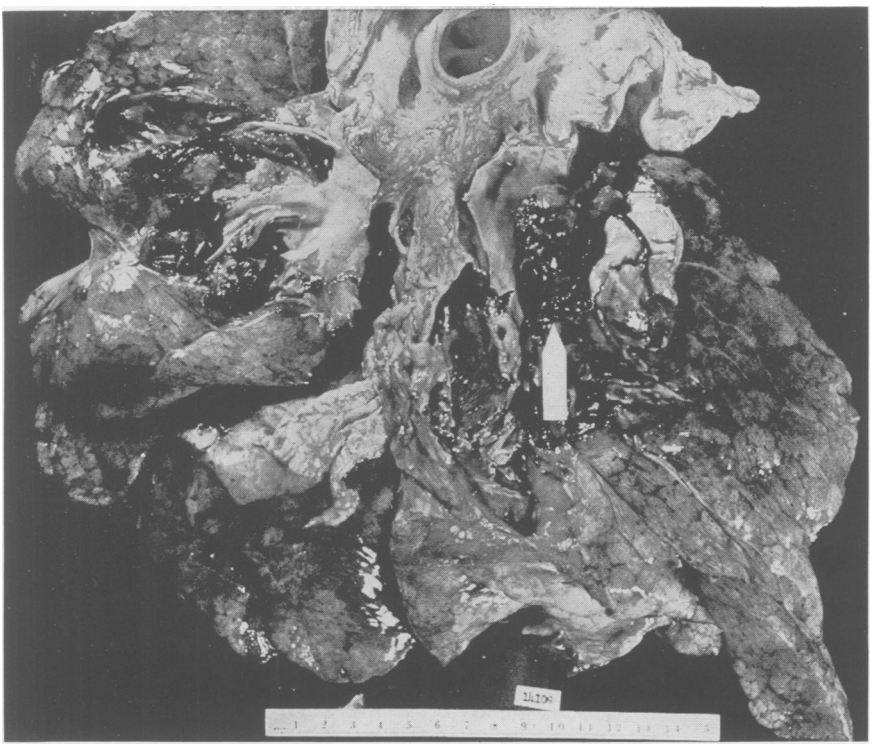


FIG. 1. Photograph at autopsy showing thrombus occluding left main pulmonary artery (Case 1).

dium succinate, was 37 seconds on one occasion and 62 seconds on another. There were no symptoms or signs of peripheral venous thrombosis. The electrocardiograms were not definitive for a myocardial infarction. The possibility of an intraperitoneal lesion was suggested by a bizarre pattern of abdominal pain and tenderness. Circulatory collapse and death occurred 11 days after the onset of dyspnea, and 24 days after thyroidectomy.

At autopsy there was an adherent thrombus, measuring 4.0 by 4.5 cm., in the left main pulmonary artery, extending into the smaller arterial branches. There were no thrombi in the main pulmonary artery or the right branch (Fig. 1). Dilatation and hypertrophy of the right auricle and ventricle were present. The pelvic veins contained small thrombi.

The changes of early organization in the thrombus were compatible with the brief duration of the clinical course, and suggested that the terminal episode developed as a postoperative complication rather than being a long standing preexisting process. The clinical picture was sufficiently suggestive to have warranted angiocardiography, a procedure which had actually been discussed and which would have provided the correct diagnosis. The anatomic location of the thrombus was such as to raise the question of surgical accessibility.

This experience sensitized the authors to the real possibility of salvage, and the problem had received considerable thought and discussion before the second case was admitted to the hospital one year later.

Case 2. F. W., admission number 86935, was a 53-year-old white female who was admitted to the Montefiore Hospital on January 26, 1957, with a chief complaint of severe dyspnea and cyanosis commencing on the previous day. Following pregnancy 30 years previously there had been an episode of thrombophlebitis. Ten years prior to admission the patient was treated with digitalis, salt restriction, and diuretics for recurrent episodes of dyspnea. The blood pressure was normal, no cardiac murmurs were heard, and there was no evidence of either rheumatic heart disease or coronary arteriosclerosis. Exertional dyspnea increased, and the patient noted dilated and pulsatile neck veins. The heart was described as enlarged by fluoroscopy six years prior to admission. One year before the final episode the patient developed thrombophlebitis in both lower extremities, also

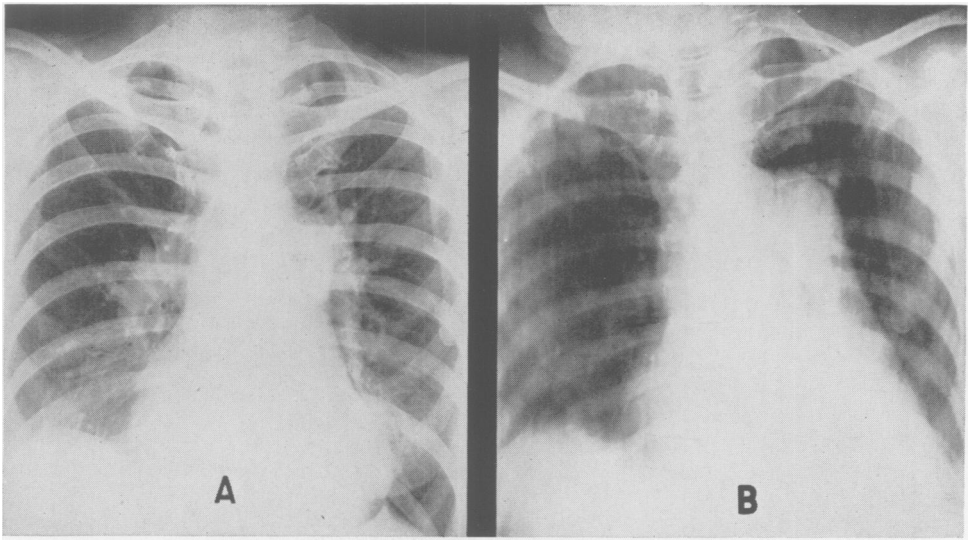


FIG. 2. Case 2. P.A. roentgenograms of chest (A) 9/30/55 and (B) 1/26/57 (portable), showing prominent pulmonary artery, increased transverse diameter of the heart, and clear lung fields.

involving the veins of the lower abdominal wall, and she was hospitalized at that time with chest pain and hemoptysis. An area of infarction in the left upper lobe was demonstrated, and this gradually cleared. The dyspnea increased in frequency and severity, and did not respond to the usual treatment for congestive heart failure. Cyanosis of the lips and nail beds was noted six weeks before admission, and had progressed markedly. Physical examination disclosed an acutely ill, severely dyspneic, markedly cyanotic white woman, sitting upright in bed. Although the blood pressure could not be obtained, and no pulse could be felt in the neck or extremities, the patient was fully conscious and alert. The neck veins were greatly distended, and pulsated. A patch of fine crackling rales was heard at the right base posteriorly. The heart was enlarged, the point of maximum impulse being 1 centimeter outside the mid-clavicular line in the fifth interspace and the left border of dullness 2 centimeters outside the mid-clavicular line. Retrosternal dullness was increased in width, with dullness to the right of the sternum. The rhythm was regular, with a rate of 96, an accentuated pulmonic second sound, and a proto-diastolic gallop at the apex. The heart sounds were distinct and of fair quality, and there were no murmurs or thrills. The liver edge was felt three fingers below the right costal margin, and the spleen was not palpable. There was no ascites, and there was slight pitting edema of both lower extremities. A positive hepatojugular reflex was present.

The positive laboratory data included a hematocrit of 55 per cent, with a white blood cell count of 8,850. There was a 3 + albuminuria, with a 1 + glycosuria and 2 to 4 leukocytes per high power field. The venous pressure was 300 mm. of water, and the decholin circulation time was measured at 70 seconds on two determinations. The electrocardiogram showed peaked P waves in leads I and II, with marked right ventricular strain pattern. A bedside roentgenogram of the chest showed considerable enlargement of the pulmonary artery segment, with an increase in the transverse diameter of the heart; there was a pleural reaction at both bases (Fig. 2).

The diagnosis of thrombosis of the pulmonary artery was made and the patient was immediately prepared for operation. Under circumstances of election, it had been planned to approach the problem of clearing the pulmonary artery of thrombus by the method currently employed in the operation for open pulmonic valvulotomy, utilizing venous inflow occlusion of the right heart and hypothermia. Pulmonary arteriotomy could be performed under these conditions with 8 to 10 minutes of a bloodless field, which should be adequate for many cases. As further evidence of the feasibility of this approach, the operation for pulmonic valvular stenosis by this technic may be readily accomplished with 2 to 3 minutes of venous inflow occlusion. However, since this patient was in extremis when placed in the sitting up position on the operating table, there was no time for the induction of hypothermia. It was planned to accomplish as much of a thrombectomy as possible

within repeated periods of 2 to 3 minutes of venous inflow occlusion under normothermic conditions, allowing the circulation to be restored for several minutes between manipulations. She was intubated promptly, and given a minimum of light anesthesia. The chest was opened by a transverse incision extending from the left to the right axilla beneath both breasts. Both pleural cavities were entered in the third interspaces anteriorly, and the sternum was divided with a Gigli saw. The pericardium was opened by a transverse incision across the base of the heart, with a vertical component anterior to the right phrenic nerve. The heart was greatly enlarged, the entire anterior surface consisting of the right ventricle, with considerable engorgement of the right auricle as well. Tapes were quickly passed around the superior vena cava, which was moderately enlarged, and the inferior vena cava, which was huge and tense, but did not appear to contain clot. The main pulmonary artery was somewhat dilated and soft. The bifurcation of the main pulmonary artery was greatly dilated and occupied by a solid mass, which extended into the right and left main pulmonary arteries as far as the roots of both lungs. Inflow occlusion was accomplished by snugging the caval tapes, an incision was made in the main pulmonary artery proximal to the thrombus (Fig. 3), and large amounts of organized clot densely adherent to the wall of the main pulmonary artery and left main pulmonary artery were scooped out. The period of occlusion had been 2½ minutes, when flow was re-established by placing a clamp across the incision in the pulmonary artery and releasing the caval occlusion. At this point the heart stopped completely, and all efforts to induce a cardiac beat were unsuccessful. Injections of calcium chloride, and adrenalin, separately and in combination, and rhymthic cardiac compression were of no avail.

It was the operator's impression that the left main pulmonary artery had been cleaned out to a considerable extent, and it had been planned to approach the right main pulmonary artery similarly if a heart beat had been reestablished. If the patient had survived the procedure, ligation of the inferior vena cava below the renal veins had also been contemplated.

At autopsy there was no intrinsic cardiac or pulmonary disease, aside from the thrombosis of the pulmonary artery and its sequalae. The residual of an old infarct was present at the left apex, and there was an organizing infarct in the left lower lobe. The main trunk of the pulmonary artery was tremendously dilated, but free of thrombus. In the right main pulmonary artery there was a dense friable organized thrombus. This was adherent to the anterior, superior, and inferior aspects of the

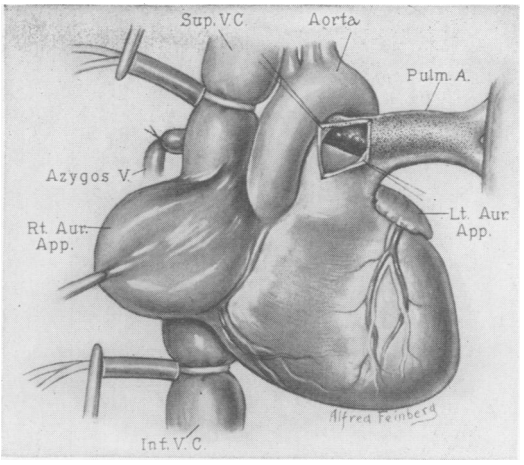


FIG. 3. Diagram of venous inflow occlusion and pulmonary arteriotomy; ligation of azygos vein may be avoided by intrapericardial occlusion of superior vena cava.

main branches, with a narrow slit posteriorly which was patent. The thrombus extended across the bifurcation to the left main pulmonary artery, but did not enter the main trunk. At the hilus it extended to the lobar branches, with involvement of a few of the segmental branches. The thrombus in the left main pulmonary artery was fragmented, large portions of it having been removed surgically (Fig. 4). Portions of the thrombus had broken off and were present in lobar segmental branches to the right middle lobe, right upper lobe, and left lower lobe. There was marked dilation and hypertrophy of the right auricle and ventricle, with widening of the pulmonary conus. A gelatinous thrombus was present in the right auricular appendage, measuring 1×2 cm. The only thrombotic focus found in the remainder of the venous system was a subintimal fibrous plaque in the left popliteal vein.

This second case is a classical example of the syndrome of chronic thrombosis of the pulmonary artery. It is believed that this could have been recognized as a potential surgical condition many weeks, months, or even years earlier, had an index of suspicion been high. The diagnosis could have been confirmed by an angiocardiogram, and elective thrombectomy might have been performed under optimum conditions. The history of an antecedent episode of thrombosis of the leg veins, followed by

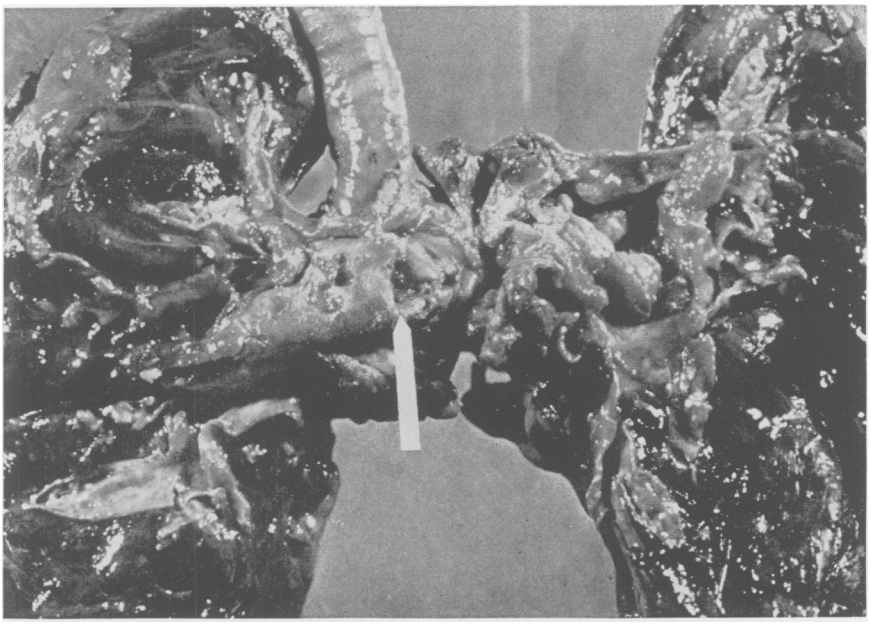


FIG. 4. Photograph at autopsy showing right pulmonary artery packed with thrombus, the left having been partially cleared at surgery (Case 2).

dyspnea unrelated to any other cardiac or pulmonary pathology, should alert one to this mechanism as the etiologic factor. It has been shown in the experimental animal that the pulmonary arterial lumen must be narrowed to approximately one-third of the normal caliber before significant evidence of obstruction appears.[20] Apparently partial obstruction within the pulmonary arterial tree can be tolerated for a prolonged period of time. During this interval, the degree of obstruction may be augmented either by the addition of subsequent emboli, or by local thrombosis. The details of the differential diagnosis of cor pulmonale due to chronic obstruction of the pulmonary artery are discussed at length in a current analysis of the problem.[1] Mitral stenosis, pulmonic valvular stenosis, auricular septal defect, acute pulmonary infarction, constrictive pericarditis, idiopathic pulmonary hypertension, and chronic pulmonary fibrosis are among the more common conditions that must be considered.

Given the opportunity to operate for chronic thrombosis of the pulmonary artery under elective conditions, the major factor influencing a successful outcome will be the location and extent of the thrombotic material within the pulmonary arterial tree. It is apparent in Table 1 that a substantial number of these cases should be amenable to simple thrombectomy by direct pulmonary arteriotomy. The operation described in this report would seem to be the first planned surgical attempt to perform thrombectomy for chronic thrombosis of the pulmonary artery. There are several additional cases in Table 1 in which the ramifications of the propagating thrombus could only be removed by incising or dividing the main or lobar arteries at the hilus. This is feasible by shutting off the flow of blood to only one lung at a time, preferably doing the worst side first. By this technic it might be possible to extract the thrombi extending toward the lobular arterial branches. Because there are cases in which it would be impossible to reestablish flow through the main pulmonary artery or its main right or left branches, an experimental program was instituted in which the main pulmo-

nary artery and the area of the bifurcation were replaced by aortic homografts (see below).

The status of inferior vena caval ligation and of anticoagulation in relation to this problem is not completely clear at this point. With a definite history of phlebothrombosis or thrombophlebitis, it would seem the part of good prophylaxis to ligate the inferior vena cava below the level of the renal veins. Since most cases of chronic thrombosis of the pulmonary artery originate as emboli from the pelvis or lower extremities, one need not require a positive history as the definitive indication. On the other hand, since one would almost certainly place a patient on anticoagulant therapy following successful pulmonary arterial thrombectomy, it might be wiser to defer the caval interruption.

Acute Embolization to the Pulmonary Artery

In 1908, Trendelenburg reported the first surgical attempts to save the lives of patients who had sustained massive acute embolization to the pulmonary artery. Through a vertical incision to the left of the sternum, with a horizontal extension over the second rib, the medial portion of which was resected, he opened the pericardium and passed a rubber tape beneath the aorta and pulmonary artery. With compression on the tape, he incised the pulmonary artery, extracted the clot, closed the pulmonary arteriotomy with a clamp, and removed the compression from the great vessels, within a period of 45 seconds. He was then able to repair the pulmonary arteriotomy without undue haste. Considering the facilities at his disposal at that time, it is truly remarkable that both of these cases were alive at the end of the operations. The first died 15 hours later in heart failure, and the second succumbed to hemorrhage from the internal mammary artery 37 hours after operation.[21]

Trendelenburg made use of the observation that many fatal instances of acute massive pulmonary arterial embolization are characterized by survival for several hours before the demise of the patient. During this comparatively short interval there is ample time to perform embolectomy of the pulmonary artery. A precise preoperative diagnosis may be established by angiocardiography. It is proposed that the technic of bilateral transsternal thoracotomy, as employed in the case of chronic pulmonary artery thrombosis in this communication, should be utilized also for cases of acute pulmonary artery embolization. The procedure may be carried out quickly, the exposure is magnificent, a bloodless field may be achieved quickly by venous inflow occlusion, and the arteriotomy and embolectomy performed with dispatch. Hypothermia should not even be necessary in these acute cases, since in all probability the period of inflow status could be kept well below the three minutes currently considered to be compatible with safety in terms of cerebral anoxia.

Congenital Stenoses of the Pulmonary Artery

In 1954, Sondergaard described three cyanotic patients in whom a marked constriction was found in the region of the bifurcation of the pulmonary artery, with a normal main pulmonary artery. He proposed the name of coarctation of the pulmonary artery for this condition.[22] A similar case had been described in 1953 by Shumacker and Lurie; through a cardiotomy they dilated the stenotic calcific ring, and described improvement in the condition of the patient.[23] Arvidsson and his associates in 1955 described 4 cases in which stenosis of the main or smaller branches of the pulmonary artery was demonstrated as a cause of pulmonary hypertension by the use of selective angiocardiography.[24] Kjellberg *et al.* described one case,[25] and Gyl-

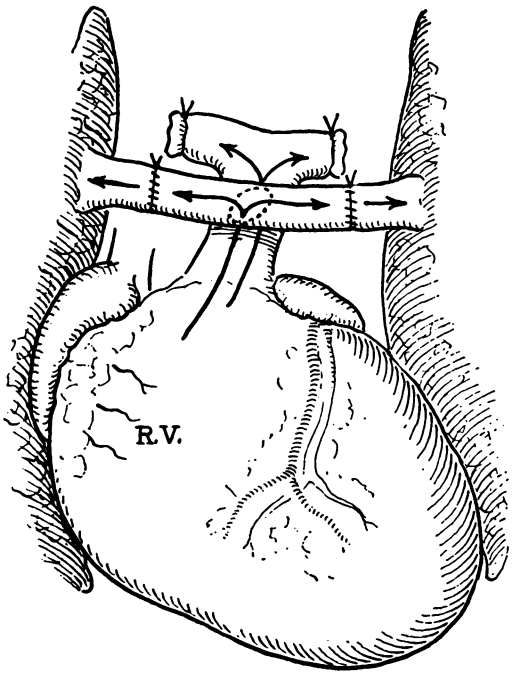


FIG. 5. Diagram of replacement of main pulmonary artery and area of bifurcation by an aortic homograft.

lenswärd and co-workers eight cases of multiple stenoses of the pulmonary artery and its branches.[26] The cases were diagnosed by means of cardiac catheterization and selective angiocardiography; the film reproductions in the last paper are exquisite.

The problem raised by some of these cases, some of the cases of chronic thrombosis of the pulmonary artery, and other situations such as aneurysms of the pulmonary artery, is that of definitive replacement of the main pulmonary artery and the region of the bifurcation, possibly extending to include some of the lobar branches. An experimental program has been instituted in the laboratory to find a solution to this challenge. Originally it was anticipated that cardiopulmonary bypass and elective cardiac arrest would be necessary, and a number of dogs were operated on with replacement of the pulmonary artery by an aortic homograft, utilizing a bubble oxygenator for the bypass and potassium citrate for the cardiac arrest. It quickly became apparent that these technics were not essential, and, in fact, not only complicated the procedure unnecessarily, but were also attended by a high mortality. It has been possible consistently to replace the main pulmonary artery and the area of the bifurcation with an aortic homograft in dogs, employing the principle of a side-to-side anastomosis between the graft and the anterior wall of the main pulmonary artery, and end-to-end anastomoses at both lung roots (Fig. 5). Since only one lung is deprived of pulmonary arterial blood at a time by this technique, there is no need for an artificial oxygenator. A preliminary report of these investigations is in preparation,[27] and dogs are being maintained on a survival basis for long term observations.

Summary

1. Chronic thrombotic obstruction of the pulmonary artery may be present for a substantial period of time, before resulting in death due to mechanical obstruction to the flow of blood from the right side of the heart. During this period a characteristic clinical picture is present which may be amenable both to accurate diagnosis and to surgical correction.

2. Two cases of chronic thrombosis of the pulmonary artery have been described, and surgical thrombectomy attempted in one patient who had exhausted her cardiac reserve.

3. Although this was unsuccessful, it should be a feasible operation when performed as an elective procedure during the interval between the development of symptoms and death of the patient.

4. The technic of the operation, with vena caval cardiac inflow occlusion, is described; the addition of hypothermia is discusssed.

5. The application of this technic to the treatment of acute pulmonary artery embolization is recommended.

6. Replacement of the main pulmonary artery and the area of the bifurcation by an aortic homograft has been accomplished successfully in dogs, and may be applicable to selected cases of congenital stenoses of the pulmonary artery and extensive cases of thrombosis of the pulmonary artery.

References

1. Schein, C. J., H. Rifkin, E. S. Hurwitt and A. Lebendiger: The Clinical and Surgical Aspects of Chronic Pulmonary Artery Thrombosis. Arch. Int. Med. In press.
2. Ball, K. P., J. F. Goodwin and C. V. Harrison: Massive Thrombotic Occlusion of the Large Pulmonary Arteries. Circulation, **14**:766, 1956.
3. Belt, T. H.: Late Sequelae of Pulmonary Embolism. Lancet, **2**:730, 1939.
4. Billings, F. T.: Primary Thrombosis of the Pulmonary Artery. Penn. Med. J., **25**:152, 1921.
5. Carroll, D.: Chronic Obstruction of the Major Pulmonary Arteries. Am. J. Med., **9**:175, 1950.
6. Covey, G. W.: Chronic Cor Pulmonale with Report of a Case. Ann. Int. Med., **18**:851, 1943.
7. Davison, P. H., G. H. Armitage and D. J. S. McIver: Chronic Cor Pulmonale Due to Silent Pulmonary Embolism. Lancet, **2**:224, 1956.
8. Desclin, L.: Uber Chronische Thrombosen des Hauptstammes und der Hauptäste der Arteria Pulmonalis. Frankfurt Ztschr. f. Path., **40**:161, 1930.
9. Fowler, W. M.: Obliterating Thrombosis of the Pulmonary Arteries. Ann. Int. Med., **7**: 1101, 1934.
10. Goedel, A.: Zur Kenntnis der Hypertrophie des Rechten Herzens und Schwerer Kreislaufstörung Infolge Verödung der Lungenschlagaderperipherie. Virchow's Arch. f. Path. Anat., **277**:507, 1930.
11. Hollister, L. and V. L. Cull: The Syndrome of Chronic Thrombosis of the Major Pulmonary Arteries. Am. J. Med., **21**:312, 1956.
12. Keating, D. R., J. N. Bunboy, H. K. Hellerstein and H. Feit: Chronic Massive Thrombosis of the Pulmonary Arteries. A Report of Seven Cases with Clinical and Necropsy Studies. Am. J. Roentgenol., **69**:208, 1953.
13. Ljungdahl, M.: Gibt es eine Chronische Embolisierung der Lungenarterie. Deutsch. Arch. f. Klin. Med., **160**:1, 1928.
14. Magidson, O. and G. Jacobson: Thrombosis of the Main Pulmonary Arteries. Brit. Heart J., **17**:207, 1955.
15. Mönckeburg, J. G.: Uber die Genuine Arteriosklerose der Lungenarterie. Deutsch. med. Wchnshr., **33**:1243, 1907.
16. Montgomery, G. L.: A Case of Pulmonary Artery Thrombosis with Ayerza's Syndrome. J. Path. and Bact., **41**:221, 1935.
17. Owen, W. R., W. A. Thomas, B. Castleman and E. F. Bland: Unrecognized Emboli to the Lungs with Subsequent Cor Pulmonale. New England J. Med., **249**:919, 1953.
18. Ring, A. and J. R. Bakke: Chronic Massive Pulmonary Artery Thrombosis. Ann. Int. Med., **43**:781, 1955.
19. Schneider, H. W. and H. S. Van Ordstrand: Thrombosis of Pulmonary Arteries. Cleveland Clin. Quart., **7**:284, 1940.
20. Gibbon, J. H., Jr., M. Hopkinson and E. D. Churchill: Changes in Circulation Produced by Gradual Occlusion of the Pulmonary Artery. J. Clin. Invest., **11**:543, 1932.
21. Trendelenburg, F.: Uber die Operative Behandlung der Embolie der Lungenarterie. Verhand. der Deutsch. Gesell. f. Chir., **37**: II, 89, 1908.
22. DeTakats, G., W. C. Beck and G. K. Fenn: Pulmonary Embolism. An Experimental and Clinical Study. Surgery, **6**:339, 1939.
23. Sondergaard, T.: Coarctation of the Pulmonary Artery. Dan. M. Bull., **1**:46, 1954.
24. Shumacker, H. B., Jr. and P. R. Lurie: Pulmonary Valvulotomy. J. Thoracic Surg., **25**: 173, 1953.
25. Arvidsson, H., J. Karnell and T. Moller: Multiple Stenoses of the Pulmonary Arteries Associated with Pulmonary Hypertension Diagnosed by Selective Angiocardiography. Acta. Radiol., **44**: 209,1955.
26. Kjellberg, S. R., E. Mannheimer, V. Rudhe and B. Jonsson: Diagnosis of Congenital Heart Disease. Chicago, Year Book Pub., 1955.
27. Gyllenswärd, A., H. Lodin, A. Lundberg and T. Möller: Congenital, Multiple Peripheral Stenoses of the Pulmonary Artery. Pediatrics, **19**:399, 1956.
28. Hurwitt, E. S., G. Robinson, P. Glotzer and M. Gilbert: Replacement of the Main Pulmonary Artery and Its Bifurcation by an Aortic Homograft. In press.