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AORTO-PULMONARY SEPTAL DEFECT WITH PATENT DUCTUS ARTERIOSUS AND DEATH DUE TO RUPTURE OF DISSECTING ANEURYSM OF THE PULMONARY ARTERY INTO THE PERICARDIUM

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Aorto-pulmonary septal defect, which has been regarded as a rare congenital cardiac lesion, was first reported by Elliotson in 1830. A number of single examples were recorded near the beginning of the present century, but the condition was then little mentioned until recent years. In 1936 Maude Abbott included 10 in her series of 1,000 cases of congenital heart disease. Dadds and Hoyle (1949), Spencer and Dworken (1950), Gasul, Fell, and Casas (1951) have fully reviewed the previously reported cases. The last writers found 24 such lesions, but in the discussion following the paper of Scott and Sabiston (1953) various speakers mentioned personal experience, giving the impression that the condition was more common than had previously been thought. This recent increase in the discovery of aorto-pulmonary septal defects is probably due to the extension of cardiac surgery, and to the importance of distinguishing such defects from patent ductus arteriosus. In the past many have undoubtedly passed as patent ductus. Gibson, Potts, and Langewisch (1950) report four which were so diagnosed and found at operation to have aorto-pulmonary septal defects which were not treated. Scott and Sabiston (1953) report the successful surgical treatment of one, as well as considerable work on the production and the surgical treatment of such defects in dogs. Cohen, Warden, and Lillehei (1955) mention a patient who died at operation and describe work on the production and repair of such defects. Gross (1952) and Fletcher, DuShane, Kirklin, and Wood (1954) report single examples which have been successfully repaired.

The majority of the defects are of small or moderate size, being less than 10 mm. in diameter and giving rise to signs and haemodynamic effects similar to those of patent ductus arteriosus. There are notable exceptions where the defect is much larger. Dadds and Hoyle (1949) and Bain and Parkinson (1943) each report a defect which was over 5 cm. in the least diameter. The case here reported is of this order and is notable for the long time of survival (26 years) in spite of the large defect. Dadds and Hoyle found that the average age of death in the previously reported series was 14 years, this figure being greatly influenced by the survival of two patients with small defects who lived for 37 and 48 years respectively. Their own patient died at the age of 16 years and that of Bain and Parkinson at the age of 18 years. Bailey (1955) also points out that the prognosis is much poorer than with patent ductus arteriosus, probably due to the larger calibre of the defect and the greater shunt involved. This may also account for the fact that bacterial endocarditis occurs so rarely in association with the defect. The only record of this complication is made by Cohen and others (1955), who mention that in their case which did not survive operation bacterial endocarditis had been successfully treated by antibiotics five years previously.

In the more severe examples the amount of shunting from the aorta to the pulmonary artery must be limited by elevation of the pulmonary artery pressure above normal level, due either to persistence of the foetal pulmonary vascular resistance (Hultgren, Selzer, Purdy, Holman, and Gerbode, 1953) or to the development of an increased resistance. This mechanism commonly exceeds the bounds of necessity as shown by the reversal of the shunt with the appearance of terminal cyanosis in many. This event is usually quickly followed by death in uncontrollable congestive failure. The present case is notable for the long duration of the central cyanosis (five years) and the marked finger clubbing, which is usually absent.

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In reported cases it is unusual to find other congenital cardiac defects. Dadds and Hoyle have reviewed the literature, and the only instance of coexisting patent ductus arteriosus was reported by Hektoen (1900) in a newborn infant in whom the foramen ovale was also widely patent. In the present patient the ductus was definitely patent (internal diameter 5 mm.), but could have had little haemodynamic significance beside such a large additional defect.

No case has been reported in which death was due to rupture of the pulmonary artery.

Case History

A man, (F. W.) aged 26 years, was single and a baker's confectioner by occupation. The family history and the previous history of serious illness were both negative. Both the patient and his parents were of low intelligence and gave poor histories. His mother reported that he was the first-born of four children. The pregnancy was normal, but he was born four weeks prematurely and was quiet and backward as a baby. At the age of 2 years he was unwell, and the attending doctor told the parents that he suffered from congenital heart disease. Apart from frequent severe nose bleeds and the occasional vomiting of clotted blood he remained well and developed reasonably normally. The symptoms of bleeding ceased at the age of about 14 years.

The patient reported that ever since he could remember he had been more breathless on exertion than his fellows and did not play games at school. At the age of 15 years it was noticed that he became cyanosed after severe exertion, and at the age of 21 years, or five years before his death, he was noted to be permanently and progressively cyanosed.

He remained well, leading a normal life and doing light work, until September, 1954, when rapidly progressive breathlessness forced him to stop work. At that time he was dyspnoeic on walking for even a short distance on the flat. In December of the same year he developed widespread oedema which re-

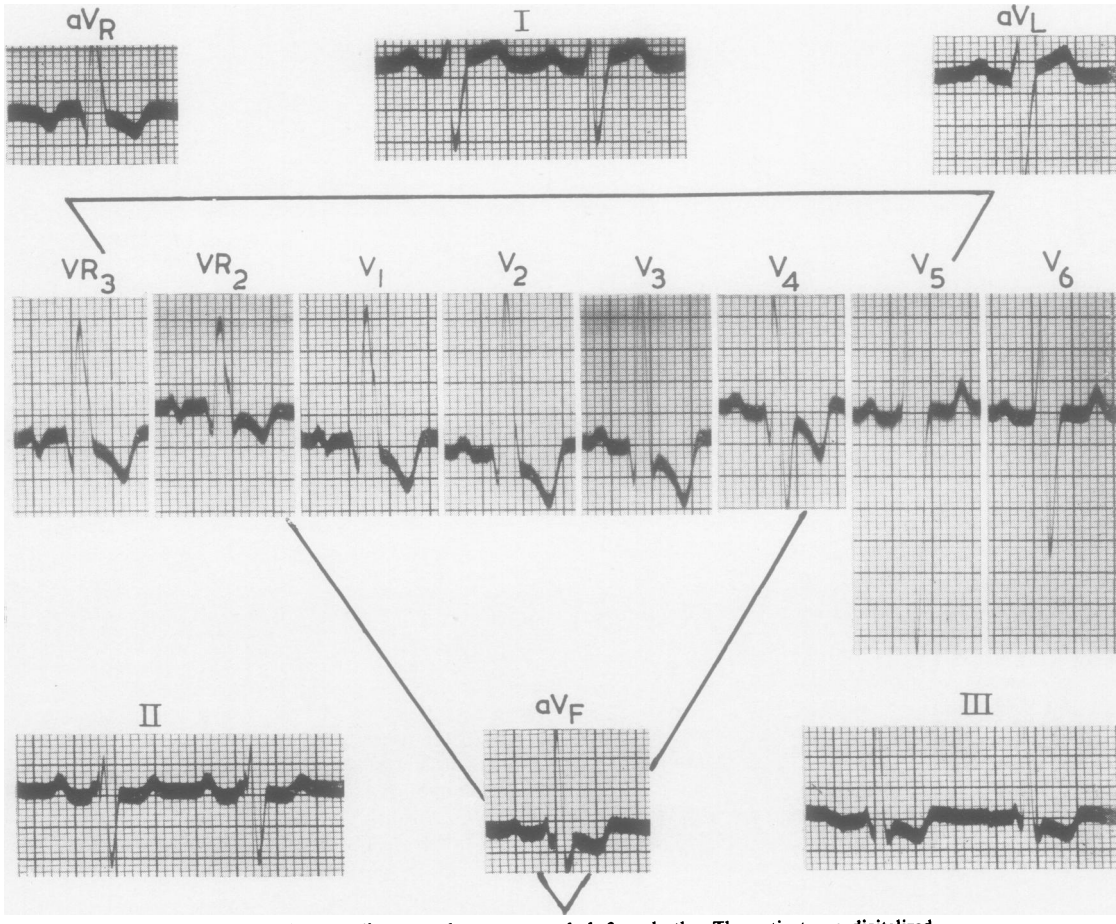

Fig. 1.—Electrocardiogram taken one month before death. The patient was digitalized.

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sponded well to rest, salt restriction, digitalis, and mercurial diuretics. He was allowed to get up and was referred to Sully Hospital for investigation in March, 1955. At that time he was able to walk 200 yards on the flat without dyspnoea, he was not orthopnoeic, and had no paroxysmal nocturnal dyspnoea or other symptoms.

On examination he was seen to have gross central cyanosis and there was marked clubbing of the fingers and the toes. The pulse was regular and of full volume and all pulses were normally palpable. The blood pressure was 130/75 mm. Hg in both arms. The jugular venous pressure was elevated to 6 cm. above the sternal angle with a dominant "a" wave. There was no oedema. The anterior chest wall bulged over the praecordium, there was a very forceful right ventricular thrust and a marked lift over the pulmonary artery felt maximally far laterally in the second left intercostal space. The pulmonary second sound was readily palpable. On auscultation in the tricuspid area there was a presystolic gallop. A marked systolic ejection click was heard maximally in the pulmonary area; there was a systolic murmur (with thrill) and wide splitting of the second heart sound with very marked accentuation of the pulmonary element. There were no other murmurs. The liver was moderately enlarged and firm. There was no oedema.

The electrocardiogram (Fig. 1) showed a vertical heart, a prominent P wave with marked right ventricular hypertrophy, and a degree of intraventricular block (QRS− 0.14 second). A 6-ft. radiograph (Fig. 2) and fluoroscopy showed the lung fields to be oligaemic peripherally and there was aneurysmal dilatation of the pulmonary artery showing little pulsation. There was gross general cardiac enlargement, but especially of the right heart. The haemoglobin was 16.28 g.% (110%) and full blood examination, including enumeration of platelets and bleeding and clotting times, was otherwise normal.

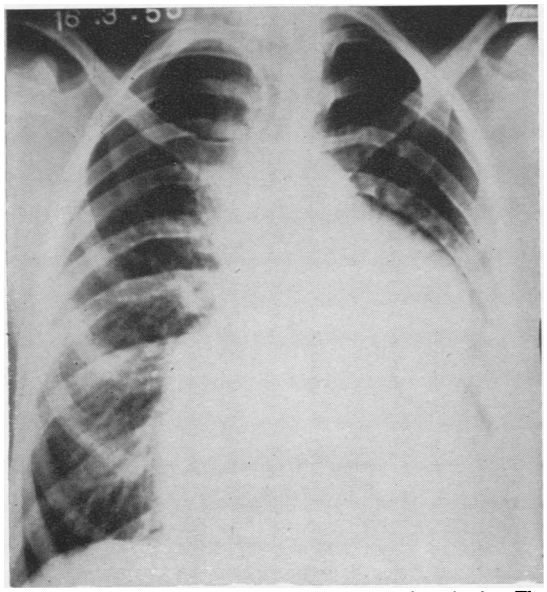

FIG. 2.—Postero-anterior radiograph a month before death. The general cardiac enlargement and the aneurysmal dilatation of the pulmonary artery are well seen. The lack of vascularity in the periphery of the lung fields is most obvious in the upper lobes in this reproduction.

At the first attempt at cardiac catheterization the extreme size of the right heart made it impossible to traverse the pulmonary valve. The findings in the right atrium and ventricle were identical with those made subsequently (Table I). On April 19, 1955, catheterization was repeated using a No. 8 F catheter of 125 cm. length. This was successfully passed through the pulmonary valve and the pressures shown in Table I recorded. The pressure in the pulmonary artery was thus considerably higher than that in the brachial artery.

TABLE I

PRESSURES RECORDED AT CARDIAC CATHETERIZATION

Chamber	Pressure (mm. Hg) above Sternal Angle
Right atrium	"a" wave=6 "v" wave=2 Mean=zero
Right ventricle	125/0
Pulmonary artery	125/62 Mean=70
Brachial artery	90/75

(Pressure waves were recorded using a Sanborn electromanometer and "polyviso" direct writing recorder.)

The extreme density of the pulmonary artery shadow made it very difficult to see the catheter in that site. On attempting to pass it further, the loop in the right ventricle immediately set up runs of ventricular ectopic beats, and so the catheter was withdrawn. No blood samples were studied. Catheterization had confirmed the clinical impression that this was a case of the Eisenmenger syndrome type with pulmonary hypertension and right to left shunting, though the situation of the shunt was not certainly known. As no operation could be helpful, it was felt that it was not justifiable to subject him to the risk of angiocardiography.

Following catheterization his condition was good, and on April 25, preparatory to discharge, four carious teeth were removed under local anaesthesia and with penicillin cover. This was followed by an insignificant amount of bleeding, but five hours later he suddenly collapsed and died, making no response to resuscitative measures. Necropsy was performed by Dr. R. M. E. Seal. The relevant findings only are recorded.

The pericardium was distended by over a litre of partly clotted blood which came from a tear at the base of the pulmonary artery (Fig. 3). The heart was grossly enlarged, weighing 900 g. The right ventricle was enormously dilated and hypertrophied, its average thickness being 1.3 cm., while in places it was 2.5 cm. The left ventricle was also dilated and hypertrophied up to 2.2 cm. The pulmonary artery was

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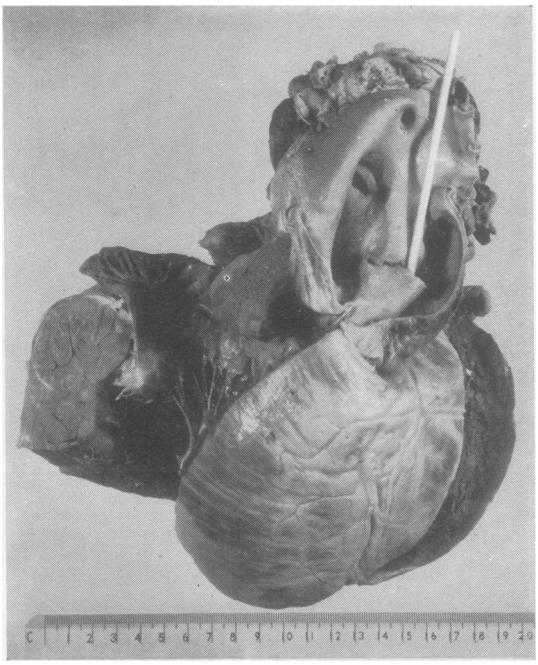

FIG. 3.—Fixed specimen of the heart and great vessels, looking into the right ventricle. Note the size of the ventricles and the thickness of the walls. The normal tricuspid and pulmonary valves can be seen. The aneurysmal pulmonary artery has been cut back and everted for demonstration purposes. The glass rod lies in the tear in the pulmonary artery and shows the direction of the dissection. The aorto-pulmonary septal defect is to the left of the tear and the opening of the right pulmonary artery to the right of the rod.

greatly dilated and showed considerable thickening of the wall. Between the pulmonary artery and the aorta, and beginning 1 cm. distal to the valves, was a large oval defect measuring 3 cm. × 4.5 cm. with its long axis in the length of the vessel; 3 cm. distal to the upper end of this was the patent ductus arteriosus, measuring 5 mm. in internal diameter. The pulmonary and aortic valves were both normal and competent. In the grossly dilated pulmonary artery was a transverse tear 3.5 cm. in length (Fig. 4), which, after a short dissection within the wall, led into the pericardium. There was no more than slight atherosclerosis of the pulmonary artery. The other organs showed the changes of chronic venous congestion. The liver was enlarged and presented a remarkable nobbly surface. On section there were innumerable pale, circumscribed areas many of which were a centimetre or more in diameter and separated by coarse strands of dense fibrosis. Macroscopically the appearances were suggestive of multiple foci of hepatoma formation, but on section were shown to be hyperplastic regenerative nodules in an organ severely affected by cardiac cirrhosis.

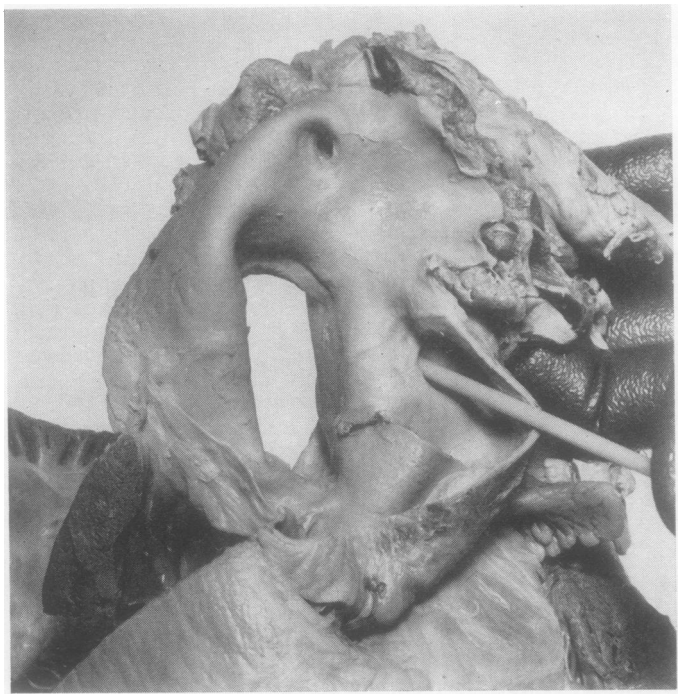

FIG. 4.—Closer view of the pulmonary artery which is further everted and rotated to demonstrate the opening of the defect looking through into the aorta. The patent ductus lies immediately above the defect. The glass rod is in the right pulmonary artery, and where it crosses the cut wall of the aneurysmal portion of the pulmonary artery the thickness of that vessel can be seen. The clean edge of the tear is also visible.

Microscopy also showed that the main pulmonary artery was 50% thicker than the aorta at the same level. The dissection had occurred in the media between the outer third and the inner two-thirds, splitting it cleanly. Compared with the aorta there was much less muscle, the elastic fibres were sparse and coarse, and there was a general increase in the collagen. There were many cystic areas of mucoid degeneration containing faintly blue-staining material, which frequently showed cleft formation, and sometimes the spaces were filled with red cells.

The muscular pulmonary arteries showed no more than moderate medial thickening and there was only occasional slight intimal fibrosis.

DISCUSSION

EMBRYOLOGY.—Shepherd, Park, and Kitchell (1944) suggest that the defect is established between the fifth and eighth weeks of foetal life, during which time the septum in the truncus arteriosus is forming and the great vessels are rotating. Extreme cases of aorto-pulmonary

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septal defect may resemble persistent truncus arteriosus, but, as Bain and Parkinson (1943) pointed out, there is no defect of the interventricular septum or of the pulmonary or aortic valves. The defect is usually a centimetre or less above the pulmonary valves and thus distant from the site of the ductus. This is clearly illustrated in the present case (Fig. 4). Care must be taken to distinguish these lesions from acquired rupture of an aneurysm of the sinus of Valsalva, where the communication is below the level of the pulmonary valves.

DIAGNOSIS.—Previous papers have discussed at length various clinical features which may enable this condition to be diagnosed and, most important, to be distinguished from patent ductus arteriosus. Among the points made are the "atypical" nature of the machinery murmur, the absence of, or lack of, rumbling quality in the diastolic murmur, the maximum intensity of the murmur in the third rather than the second left intercostal space and its tendency to be louder nearer the mid-line than is customary in patent ductus arteriosus. In the present case these points were of little help, as the auscultatory signs were simply those of extreme pulmonary hypertension, though it is not usual to get a systolic thrill from this cause alone. From previous papers it is evident that there is no certain clinical method of establishing the diagnosis and the best that can be expected is to suspect the condition and then investigate further by other methods.

Electrocardiography and fluoroscopy are not of great help except in cases of large defects where the extreme size of the pulmonary artery (Fig. 2) would be unlikely to be found in patent ductus arteriosus. It is generally agreed that cardiac catheterization is not particularly helpful, but Himmelstein (1953) suggests that, when a catheter passes from the pulmonary artery to the aorta by way of an aorto-pulmonary septal defect, it then follows the course of the aortic arch to enter the descending aorta. However, when traversing a patent ductus it promptly enters the descending aorta without traversing the arch. In a case such as that under discussion, quite apart from the fact that both lesions were present, the extreme size and radio-opacity of the great vessels made it impossible to see the course of the catheter. In lesser degrees of defect, where the diagnosis is most important, this may be helpful.

Venous angiocardiography has not been successful in differentiating aorto-pulmonary defects from patent ductus arteriosus. Gasul and others (1951) have successfully outlined the defect in two cases by the use of retrograde angiography, and Scott and Sabiston (1953) have also used this method to establish the diagnosis before operation. Retrograde angiography would appear to be an important method of investigation.

TREATMENT.—When the diagnosis has been established surgical treatment must be seriously considered. In view of the poor prognosis this should probably be carried out in childhood. Scott and Sabiston's successful case was operated on at the age of 10 months. In another aged 18 months the defect was 10 mm. in diameter and was too large to be closed. Undoubtedly the present lesion would have been of the same type.

Where there is no prospect of surgical treatment the usual medical measures must be used. It is probably wise to place some limitation on exertion, and in fact most patients find that their dyspnoea prevents them at an early age from severe exertion. When congestive failure develops it is treated in the usual way, and the response as in this case may at first be good, but it is usually disappointing and temporary only, and death soon follows. This is the usual mode of termination in this condition.

UNUSUAL FEATURES OF THE PRESENT CASE.—Bleeding is mentioned, as severe epistaxis is also recorded by Dadds and Hoyle (1949). They found no explanation. In my patient the blood and its clotting mechanism were fully investigated and found to be normal, but this was 11 years after the excessive bleeding had ceased. As central cyanosis and venous congestion had developed in this period without the recurrence of epistaxis it is unlikely that mechanical factors within the vessels were important in its production. No explanation of this symptom is offered and its occurrence in two cases may be fortuitous.

Clubbing and Cyanosis.—The rare occurrence of clubbing is almost certainly due to the fact that the appearance of cyanosis is usually a terminal feature. In this case severe and permanent cyanosis had been present for five years before death and cyanosis on exertion for six years before that. The gross degree of clubbing is probably related to the severity and duration of the cyanosis.

Long Survival.—It is unusual for a patient with this defect to survive past the teens and those which do so have small defects. In spite of his large defect and the patent ductus, this man was holding a job and leading a normal life until a few months before his death. His cyanosis over many years was proof that the right ventricle was working against a higher pressure than the left, and this was further confirmed by the catheter finding and the

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fact that at necropsy the myocardium of the right ventricle was thicker than that of the left. The right ventricular muscle must have been more than usually resistant to failure, though the remarkable degree of change in the liver is evidence that this organ must have been congested, at least periodically, over a long period.

The Patent Ductus Arteriosus.—This is probably of academic importance only in this case, but its presence demonstrates that these two conditions do occur together and the possibility should be considered in cases coming to surgery. Its presence illustrates clearly the relative situations of the two lesions (Figs. 3 and 4).

Pulmonary Vascular Changes.—The increase in the thickness of the walls of the muscular pulmonary arterioles was of a surprisingly minor degree especially when the extreme pulmonary hypertension and its duration of many years are considered. The changes were of nothing like the degree discussed in Eisenmenger's complex by Edwards, Dry, Parker, Burchell, Wood, and Bulbulian (1954) in very much younger patients, and it would appear that much of the vascular resistance in the pulmonary circulation during life must have been provided by functional changes in the vessels with no organic counterpart. Goodwin, Hunter, Cleland, Davies, and Steiner (1955) have discussed this finding in mitral valve disease, but regard the purely functional changes as being initial only. It thus remains strange that there should be so little organic change in the vessels in this instance.

Mode of Death.—The rupture of the aneurysmal pulmonary artery with dissection down the wall and rupture again into the pericardium must be a very rare occurrence. This had many of the features of the ordinary dissecting aneurysm of the aorta and was not a simple rupture of a distended vessel. Microscopically all the features of Erdheim's cystic medio-necrosis commonly regarded as an important aetiological factor in dissecting aneurysm of the aorta (Anderson, 1948) were found in the pulmonary artery, and it is reasonable to regard the mechanism of rupture as being the same. Any possible relationship between the rupture and the cardiac catheterization was considered but was thought to be unlikely. (1) The site of the rupture was a most unlikely site for the tip of the catheter to impact. (2) The catheter was never used forcefully and indeed could not have been, for as soon as the least forward movement was made at the butt end the loop in the right ventricle touched the wall of that chamber and caused such arrhythmia that the catheter was immediately withdrawn. (3) There was no sign of haematoma or inflammatory changes at or near the rupture.

It is also unlikely that there was any important relationship to the dental operation, which was expertly done and caused little upset to the patient. It is always possible that straining during the operation precipitated the beginning of the dissection of a vessel in which it could have occurred at any time.

Summary

A case is described in which there was a large aorto-pulmonary septal defect and a patent ductus arteriosus. The patient lived to the unusually late age of 26 years, being permanently cyanosed and clubbed for the last five years, and then died from haemopericardium caused by the rupture of a dissecting aneurysm of the pulmonary artery. This vessel showed Erdheim's medio-necrosis. The clinical, cardiac catheterization, and pathological findings are described, and unusual points discussed. The recent literature is reviewed and the importance of the differential diagnosis of this condition from patent ductus arteriosus when surgery is considered is emphasized. Retrograde angiography is the only method by which this can be certainly achieved.

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