EISENMENGER'S COMPLEX

BY

A. J. GLAZEBROOK

From the Department of Clinical Medicine, University of Edinburgh

Received May 8, 1943

An opening at the base of the interventricular system in association with other defects is one of the commonest of congenital cardiac lesions. The usual combination is Fallot's tetralogy, which consists of a ventricular septal defect, dextraposition of the aorta, and stenosis of the pulmonary artery, with cyanosis.

A much rarer condition is that known as the Eisenmenger complex, where dilatation of the pulmonary artery occurs instead of stenosis, together with dextra-aorta and septal defect. This condition was originally reported by Dalrymple in 1847. Eisenmenger (1897) presented the first complete study of such a case, in which the diagnosis had been made during life by his colleague, von Schrotter.

In 1927, Maud Abbott described 8 cases, 3 of which she had studied personally. According to Baumgartner and Abbott (1929) the condition can be differentiated clinically from Fallot's tetralogy by the following points :—

- (1) absence of, or only a moderate degree of clubbing and cyanosis,
- (2) the localization over the præcordium of a harsh systolic murmur at the defect, not transmitted into the vessels of the neck,
- (3) the occasional presence of a diastolic murmur of pulmonary insufficiency, and
- (4) the distinctive character of the X-ray picture.

Hoarseness and aphonia, from pressure of the huge pulmonary conus on the recurrent laryngeal nerve, sometimes occur. Pulsation of the dilated pulmonary artery may be felt, and abnormal dullness percussed at the left base. The midsternal systolic murmur of the Eisenmenger complex is transmitted downwards to the right and to the left, as well as through to the back, whereas in the Fallot tetralogy it is usually transmitted upwards into the neck vessels and through to the back.

Polycythæmia, varying from 7,000,000 to 12,000,000 red cells, is a feature of Fallot's tetralogy, but not of the Eisenmenger complex.

CASE REPORT

A male, aged 15, had been a "blue baby" at birth in 1928, and was troubled with his chest in infancy and childhood. He always had a high complexion and frequently had a bleeding nose ; he was breathless on exertion.

There was nothing of interest in the family history.

During the period 1934–1937 he was in hospital three times with an exacerbation of his chronic bronchitis, complaining of præcordial pain on exertion, a hard dry cough, and attacks of respiratory embarrassment at night. A diagnosis of congenital pulmonary stenosis was made, and in 1937 a poor view was taken of his prognosis. Arrangements were made for his reception in a hospital for incurables ; but on arrival there with his mother he created such a disturbance that he had to be taken home : he had always been a noisy and obstreperous patient.

In 1943 the lad, now 15 years old, was seen during the course of a routine follow-up. His general health had been much better since his discharge from hospital, and he now led a very active life, although inclined to be troubled by bronchitis during the winter months. He no longer suffered from epistaxis or from præcordial pain, but sometimes took a stitch in the side when he "hurried up hills."

On examination he was small for his age but powerfully built. There was a tinge of cyanosis in the cheeks and lips, but no clubbing of the fingers. The pulse was regular in time and force, the rate 80, and the wave of good volume and well-sustained. His blood pressure was 110/80.

The left side of the chest was a little more prominent than the right, and there was diffuse pulsation in the fourth, fifth, and sixth interspaces. The apex beat was felt in the sixth space, four and a half inches from the mid-sternal line. It was diffuse and forcible. A thrill was palpable at the base, the point of maximum intensity being in the third left interspace just lateral to the border of the sternum. On percussion both the right and left borders of the heart were greatly enlarged, and the dilated pulmonary artery could also be made out, the dullness in the second left interspace extending two inches from the mid-sternal line.

On auscultation the predominant feature was a systolic murmur. It was best heard at the point of maximum intensity of the thrill but was audible all over the præcordium. It was mainly conducted downwards to the right and to the left, and through to the back, but was not heard in the neck vessels. The pulmonary second sound was inaudible beneath the prolonged systolic murmur in this area, but a distinct diastolic murmur could not be distinguished. In the aortic and mitral areas the second sound was pure and closed.

No evidence of recurrent laryngeal involvement could be found. There was an undescended testicle on the right side. Blood examinations had been carried out on various occasions, the red count varying from 5.2 to 5.5 million, and the hæmoglobin from 108 to 116 per cent. The Wassermann reaction was negative.

The chief features on X-ray examination were (a) great enlargement of the right ventricle, (b) no demonstrable enlargement of the left auricle, (c) a right-sided aortic arch, displacing the œsophagus forward, and (d) prominence of the pulmonary artery (Fig. 1 and 2).

The chief features of an electrocardiogram in 1934 were a normal sinus rhythm with runs of complete dissociation, and right axis deviation (Fig. 3). A record in 1941 showed disappearance of complete dissociation. There was an increase in the height of the P waves in

FIG. 1.—Radiogram showing hypertrophied right ventricle, dilated pulmonary artery, and dextra-aorta.

FIG. 2.—Left (II) oblique position, showing forward displacement of the œsophagus by the right-sided aortic arch.

leads I and II. The QRS interval had increased from 0.06 to 0.12 sec. Inverted T waves had developed in leads II and III, and an S-T depression in lead II. The sternal lead showed depression of S-T, and a diphasic T, while there was a steep inversion of the T wave in

FIG. 3.—Cardiogram taken in 1934, showing right axis deviation and runs of complete dissociation.

FIG. 4.—Cardiogram taken in 1943, showing larger P waves, especially in lead II, and (usually) normal rhythm. On the right ; sternal C₂F above, and apical C₄F below.

FIG. 5.—Cardiogram taken in 1943, showing occasional dissociation.

the apical lead. In 1943, little further change was recorded (Fig. 4). Infrequent runs of complete dissociation could be made out (Fig. 5), but normal sinus rhythm predominated.

AETIOLOGY

According to the theory of Spitzer (1923) so enthusiastically endorsed by Maud Abbott in her monograph, the critical period of heart formation lies between the fifth and eighth weeks of embryonic life, i.e. before the cardiac septa are formed. During this time, the complex processes of torsion, involution, re-adjustment, and fusion, necessitated by the development of the heart from a simple tube to a four-chambered structure, are taking place. The appearance of the heart in the tetralogy of Fallot is highly suggestive of the relationships that would result from the uncovering of the right reptilian aorta and the obliteration of the left, in the delayed torsion of Spitzer's theory.

The dilatation of the pulmonary artery in the Eisenmenger complex has been attributed to inflammatory lesions. Thus in Blechman and Paulin's case (1922) a streptococcal endarteritis was thought to be the cause of the dilatation. In Stewart and Crawford's case (1933) the histology of the pulmonary arteries suggested an inflammatory process. The authors gave reasons for ascribing the inflammation to a syphilitic infection, but their evidence was based purely on the histological appearances. In their case they found also an old healed endocarditis of the pulmonary valve, and in addition atherosclerotic changes in the pulmonary arteries. They suggest that the syphilitic infection might have been congenital and the chief cause of the cardiac anomalies, and that the pulmonary dilatation was quite possibly due to the arteritis.

Taussig and Semans (1940) describe a case of Eisenmenger's complex in a negro, aged 7 ; a healed lesion of the aortic valve was present, the extensive fibrosis and hyalinization showing it to be of long standing : they question whether the lesion was the result of healed bacterial endocarditis, or really related to the congenital malformation.

Inflammatory changes are frequently found in association with abnormal blood streams in the heart. Thus in *Maladie de Roger* sub-acute endocarditis commonly occurs around the margins of the septal defect and upon the wall of the right ventricle opposite the defect (Abbott, 1936). Moschcowitz (1914) described active non-rheumatic vegetations in congenital heart disease, and according to Boldero and Bedford (1924), the determining factor seems to be the access of arterial blood to the right side of the heart.

The balance of evidence is in favour of the view that the inflammatory lesions in the right circulation arise primarily as a result of the congenital abnormality. That they were so marked and widespread in Stewart and Crawford's case is no doubt related to the advanced age to which this patient lived (60 years).

The pulmonary dilatation and atherosclerosis are more simply explained as a sequel of the increased pressure in the right heart. Certainly in cases of septal defect only, very great dilatation of the pulmonary arteries may occur, with subsequent pulmonary fibrosis and the production of Ayerza's disease. Such dilatation cannot take place in Fallot's tetralogy, owing to the deformity and constriction of the lower pulmonary bulbar orifice.

PROGNOSIS

The prognosis in these cases is the only point of practical clinical interest. The pulmonary stenosis of the Fallot type forces a good proportion of the venous blood to enter the overriding aorta. This causes the cyanosis, the polycythæmia, and the very marked clubbing. The combination of defects is obviously a very serious one, and the average life is $12\frac{3}{4}$ years, although Paul White's case (1929) attained 59 years.

In the Eisenmenger type of anomaly, the dilatation of the pulmonary artery does not hinder the entrance of venous blood into the lungs. The shunt normally takes place from left to right and little mixture of venous and arterial blood occurs. Clubbing and cyanosis are therefore slight or absent, and polycythæmia is not a feature.

The prognosis is rather better than in the Fallot type, and the average life is 16 years.

Stewart and Crawford's case (*loc. cit.*), in spite of hard living, reached 60 years, untroubled by his cardiac abnormality.

The lad described in this paper presents all the clinical features of the Eisenmenger complex, and the X-ray examination supports the diagnosis. Although he had a stormy childhood, much troubled by bronchitis, in recent years his health has been much better. At his present age of 15 he leads a very active existence, and refuses to consider any curtailment of it. Rising at six every morning, he does a paper round before going to school. In his spare time he takes his full share in the activities of his group of boy scouts, joining in their physical training, their rough games, and their week-end hikes.

SUMMARY

A case presenting the clinical and radiographic features of the Eisenmenger complex is described, and the ætiology and prognosis briefly discussed.

I have to thank Professor D. M. Lyon for his help and advice.

REFERENCES

- Abbott, M. (1927). *Modern Medicine*, Philadelphia, 4, 612.
— (1936). *Atlas of Congenital Heart Dis.*, New York.
Baumgartner, E. A., and Abbott, M. (1929). *Amer. J. med. Sci.*, 177, 639.
Blechman, G., and Paulin, A. (1922). *Arch. Mal Cœur*, 15, 472.
Boldero, H. E. A., and Bedford, D. E. (1924). *Lancet*, 2, 747.
Eisenmenger, V. (1897). *Z. klin. Med. Suppl.*, 32, 1.
Moschowitz, E. (1914). *Proc. New York Path. Soc.*, 14, 18.
Spitzer, A. L. (1923). *Virchows Arch.*, 243, 81.
Stewart, H. L., and Crawford, B. L. (1933). *Amer. J. Path.*, 9, 637.
Taussig, H. B., and Semans, J. H. (1940). *Johns Hopk. Hosp. Bull.*, 66, 156.
White, P., and Sprague, H. (1929). *J. Amer. med. Ass.*, 92, 787.