MITRAL STENOSIS ASSOCIATED WITH ANOMALOUS PULMONARY VENOUS DRAINAGE INTO A LEFT SUPERIOR VENA CAVA

BY

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Anomalous drainage of some or all of the pulmonary veins into the right side of the heart is a well-recognized pathological and clinical entity. Healey (1952) collected from the literature 144 cases and added three of his own. Excellent reports on the clinical features and the findings on cardiac catheterization and angiocardiology have been published by Snellen and Albers (1952), Gardner and Oram (1953), and Whitaker (1954).

I have not been able to find a report in the literature of the association of anomalous pulmonary venous drainage with mitral stenosis.

Case Report

A 42-year-old housewife had acute rheumatic fever at the age of 20. Two years later she gave birth to a child, the pregnancy and labour being uneventful. At 38 she began to experience breathlessness on exertion, and one year later was admitted to hospital for the treatment of congestive heart failure. At 41 she had an attack of right pleuritic pain with haemoptysis, and a second attack followed two months later. These were diagnosed as being due to pulmonary emboli.

At the time of admission in April, 1954, she was considerably disabled, could not walk more than a few yards without breathlessness, and could not climb stairs at all. She was very orthopnoeic and experienced frequent paroxysmal nocturnal dyspnoea.

Examination revealed a slightly built woman, breathless at rest, with mild central cyanosis and a well-marked "mitral facies." Her pulse was irregular owing to atrial fibrillation and of small amplitude. Her B.P. was 130/±80. The jugular

Postero-anterior teleradiogram showing enlargement of the heart and a prominent superior mediastinal shadow obscuring the aorta. The main pulmonary arteries and their branches are prominent.

venous pressure was raised, and the neck veins showed systolic pulsation suggestive of tricuspid incompetence. The liver was enlarged and pulsating, and there was slight sacral oedema. The apex beat was snapping in character and there was a prominent left parasternal heave. On auscultation M1 and P2 were accentuated; a well-marked opening snap was present. A grade 3/6 pansystolic murmur was heard at the lower end of the sternum and also at the apex, where a grade 1/4 mid-diastolic murmur was present. Bilateral basal crepitations were present in the lungs.

Electrocardiography revealed atrial fibrillation, right ventricular hypertrophy, and digitalis effect. A teleradiogram showed enlargement of the heart, with a prominent superior mediastinal shadow obscuring the aorta. The main pulmonary arteries and their branches were prominent (see Fig.). Fluoroscopy demonstrated enlargement of the right ventricle, right atrium, and left atrium. No pulsation was seen in the superior mediastinal shadow.

In the pre-operative period in hospital the patient suddenly developed a left hemiplegia, which was attributed to a cerebral embolus. She was given anticoagulant therapy and the paralysis disappeared in five days. The congestive heart failure was controlled on routine treatment with rest in bed, digitalis, and mercurial diuretics.

At operation on April 22 the surgeon noted a large, somewhat tortuous left superior vena cava, into which the left superior pulmonary vein was draining. The left superior vena cava joined the left innominate vein and was thus presumed to enter the right atrium via a right superior vena cava, although this could not be determined for certain. The left inferior pulmonary vein entered the left atrium normally. The drainage of the right pulmonary veins was not visualized. The pulmonary artery was enormous. Tight mitral stenosis was present, with a mobile valve which was not calcified. There was no mitral regurgitation. A moderate split, mainly of the posterior commissure, was performed.

The post-operative course was stormy, being complicated by severe and extensive surgical emphysema, congestive heart failure, and a pulmonary embolus, but eventually the patient made a good recovery. On discharge from hospital two months after the operation she was much improved and could climb 25 stairs without discomfort. Tricuspid incompetence was still noted, but there was no congestive heart failure.

One year later she remained well and could easily climb 25 stairs without discomfort. She was able to do her own shopping and most of her housework. There was no orthopnoea and no paroxysmal nocturnal dyspnoea. Examination showed the presence of tricuspid incompetence, but no congestive heart failure. The mitral diastolic murmur had disappeared. A soft mid-systolic murmur was audible at the pulmonary area. The pansystolic murmur at the lower end of the sternum and at the apex was still present, as was the opening snap.

The electrocardiographic and radiological features were unchanged. Cardiac catheterization demonstrated the features shown in the Table. The catheter persistently coiled up in

Results of Cardiac Catheterization

Site	Oxygen Content (Vols. %)	Oxygen Saturation
Right atrium (5 different levels) ..	16	83%
Superior vena cava (right) ..	16.6	86%
Right innominate vein ..	12.75	66%
Femoral artery ..	17.9	93%

the large right atrium and could not be passed through the tricuspid valve or into the inferior vena cava. The oxygen uptake was 210 ml. a minute and the pulmonary blood flow 11 litres a minute. As the systemic blood flow could not be calculated the amount of shunt is unknown, but the probability is that a large percentage of the pulmonary arterial flow was derived from pulmonary venous blood.

Discussion

Although mid-diastolic apical murmurs may be heard in cases of anomalous pulmonary venous drainage (Gardner and Oram, 1953; Whitaker, 1954), a confident diagnosis of mitral stenosis was made in this patient because of the presence of an enlarged left atrium fluoroscopically, and a well-marked opening snap clinically. In view of the grave condition of the patient it was decided to perform a mitral valvotomy before undertaking any special investigations such as cardiac catheterization. The findings at subsequent cardiac catheterization confirmed the presence of partial anomalous pulmonary venous drainage into the right atrium via the superior vena cava.

In a patient with mitral stenosis the presence of anomalous pulmonary venous drainage would probably add to the disability by increasing the pulmonary artery pressure. Relief of the mitral stenosis would be expected to result in improvement in the patient's condition, though perhaps not to the same extent as in a patient with normal pulmonary venous drainage. The result in this case has been excellent and most gratifying.

Taussig (1947), Snellen and Albers (1952), and Whitaker (1954) noted that the typical "figure-of-8" or "cottage-loaf" cardiovascular shadow was diagnostic of total anomalous pulmonary venous drainage into a left superior vena cava. Grishman *et al.* (1951) reported a case of partial anomalous pulmonary venous drainage, and the telerradiogram reproduced in their report showed the typical "figure-of-8" appearance. However, as the difference in oxygen content between blood from the right atrium and the femoral artery was only 0.5 vol.%, which might be within the range of experimental error, it is possible that their patient in fact had total pulmonary venous drainage into a left superior vena cava. The normal arterial oxygen saturation in their patient would not exclude this possibility, as a similar finding was noted in a proved case by Gardner and Oram (1953). In the case reported here, although only part of the venous drainage of the lungs was into the left superior vena cava, the typical "cottage-loaf" cardiovascular shadow was present and was explained by the additional presence of tricuspid incompetence with subsequent increased prominence of the superior vena caval shadow.

Summary

A case is reported of the association of mitral stenosis and partial anomalous pulmonary venous drainage into a left superior vena cava. An excellent result was obtained by mitral valvotomy.

The radiological appearance was suggestive of total drainage of pulmonary veins into a left superior vena cava, and was explained by the additional presence of tricuspid incompetence.

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Opuscula Medica, a new Swedish monthly journal, contains short papers and records cases of interest discussed at staff meetings of the Södersjukhuset, Stockholm. It is edited by Dr. Sixten Kallner, head of the Department of Medicine at the Södersjukhuset. At present papers are in Swedish, but it is hoped shortly to include brief summaries in other languages.

OBSERVATIONS ON MEGALOBlastic ANAEMIAS AFTER PRIMIDONE

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Since Badenoch (1954) reported "a hitherto undescribed form of megaloblastic anaemia" in two young epileptic women receiving anticonvulsants, including phenytoin sodium and phenobarbitone, a number of papers on megaloblastic anaemia during anticonvulsant therapy have appeared.

Hawkins and Meynell (1954) remarked that this would be the first instance of drug-induced megaloblastic alteration of the bone marrow. Their case was a male epileptic who had taken phenytoin sodium, 4½ gr. (0.29 g.) daily for 13 years. Folic acid succeeded after vitamin B₁₂ had failed to cure the anaemia. Phenytoin sodium was continued throughout the treatment. Chalmers and Boheimer (1954) described the case of a patient who over a period of five years had taken barbiturates, phenytoin sodium, and, for some months, primidone before the discovery of a megaloblastic anaemia. They could not come to definite conclusions about the particular drug which caused the anaemia, there having been no previous report on possible effects of primidone on blood formation. Their patient kept well on a combination of folic acid and primidone. Rhind and Varadi's (1954) and Webster's (1954) papers both dealt with megaloblastic anaemia after phenytoin sodium, the former's patients responding to folic acid, and Webster's case being successfully treated with vitamin B₁₂. Berlyne, Levene, and McGlashan (1955) published the histories of two epileptics developing megaloblastic anaemia, the first after taking phenytoin sodium and phenobarbitone, and the second after taking primidone for three months in addition to phenytoin sodium and phenobarbitone which had been prescribed over four years. The patients did well on folic acid. Ryan and Forshaw's (1955) paper dealt with three cases of megaloblastic anaemia following phenobarbitone combined with phenytoin sodium for the treatment of epilepsy. Vaishnava (1955) added two further cases of megaloblastic anaemia due to phenytoin sodium. Girdwood and Lenman's (1956) patient developed a similar anaemia while receiving primidone and phenobarbitone for epilepsy.

From this review of the literature, it appears that most cases of megaloblastic anaemia after anticonvulsants were caused by a combination of phenytoin sodium and phenobarbitone. Only three cases are reported of megaloblastic anaemia in which primidone may have played a part, and a letter from one of us (Fuld, 1955) contains the only evidence so far of the development of megaloblastic anaemia in patients who for a considerable time have received primidone only. It occurred to us while observing these patients that an opportunity offered itself to study the relationship between primidone and the development of megaloblastic anaemia.