MITRAL VALVOTOMY IN THE YOUNGER AGE-GROUPS

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Mitral stenosis represents an end stage of rheumatic carditis. It takes 5-15 years to develop after the onset of the initial attack, and usually occurs during the 2nd or 3rd 5-year period after the initial attack.[8] Thus the occurrence of tight mitral stenosis of a type suitable for operation is unusual under the age of 16 years. The usual lesion in the 1st decade and early part of the 2nd decade is mitral incompetence.[2]

There are very few reports of mitral valvotomy operations on patients under the age of 16 years. Most of the large series include patients from the age of 18 years and over.[1, 6, 9] Logan and Turner[4] have one case aged 16 years in their 100 cases. Lurie and Shumacker[5] described 3 cases aged 12, 14 and 15 years in which progressive symptoms and disability led to mitral valvotomy.

It is therefore worth recording 4 further cases between the ages of 7 and 14, where predominant mitral stenosis was diagnosed and valvotomy performed. Three of these cases were aged 14 years and one was aged 7 years.

CASE RECORDS

Case 1. G.D., a 7-year-old Bantu, was admitted to Baragwanath Hospital under Dr. E. Kahn on 28 April 1953, with a history of palpitations and cough for 6 months. One week before admission dyspnoea became severe and sweating occurred. The face, feet and lower abdomen became swollen and the child fainted 3 times at school. Six months before admission he had a fever, with pain in the right knee.

On admission he was in a state of congestive cardiac failure.

There was a prominent left parasternal heave extending out into the axilla. On auscultation in the axilla there was a loud 1st sound, a mid-diastolic rumble and an opening snap of the mitral valve. There was no systolic murmur, but medial to the apex a systolic murmur was audible. *An electrocardiogram* showed right ventricular predominance and a mitral P wave.

An *X-ray* showed an enlarged heart. The enlargement was confined to the right ventricle. The pulmonary-artery segment was prominent. There was peripheral attenuation of the pulmonary vessels suggesting pulmonary hypertension. The enlarged left auricle caused an indentation of the barium-filled oesophagus in the right anterior oblique view. The right auricle also appeared to be enlarged. (Figs. 1 and 2).

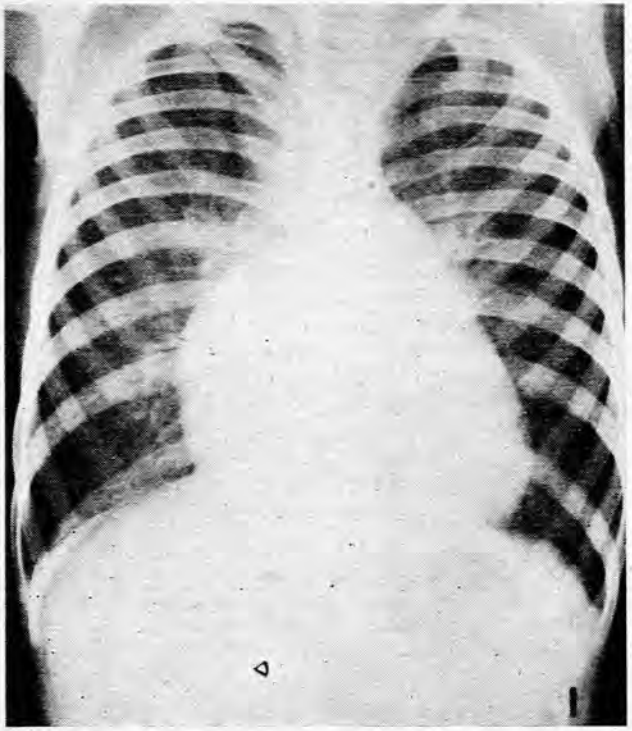

Fig. 1. Case 1: Postero-anterior teleradiogram—Showing selective enlargement of right ventricle, and prominence of pulmonary artery segment.

The child improved on bed rest and the usual treatment with digitalis and mercurial diuretics, but whenever he got out of bed he again went into failure. The erythrocyte sedimentation rate varied, but at one stage was 21 mm. in 1 hour (Westergren).

Operation. Mitral valvotomy was performed on 26 August 1953 by Mr. J. C. v. d. Spuy. A lateral approach was used. The left atrium was large and tense. The mitral valve was funnel-shaped, the orifice being one-eighth of an inch in diameter. The anterior cusp was long and mobile and the posterior cusp was short and thick. The orifice was posterior in position. The valve split easily

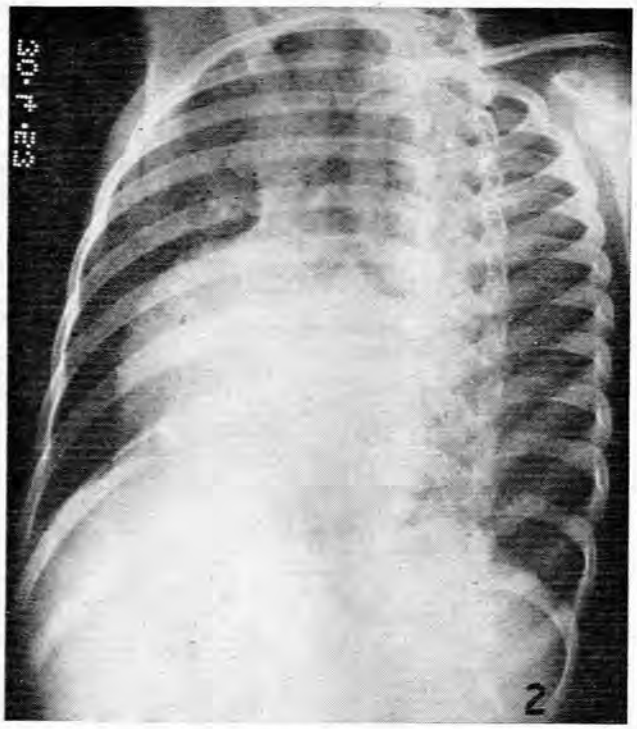

Fig. 2. Case 1: Left anterior oblique view. This plate shows the enlargement of the right ventricle.

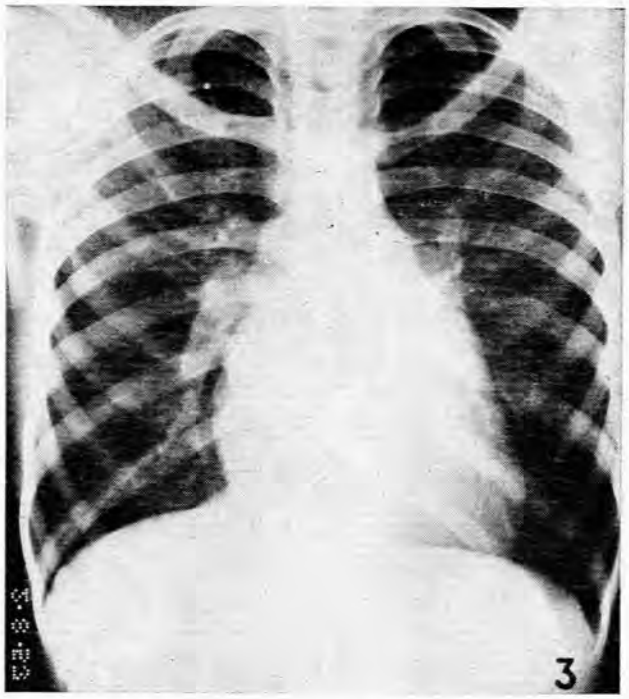

Fig. 3. Case 2: This shows a double density due to the enlarged left auricle, and a prominence of the pulmonary artery segment.

with finger fracture and the diameter of the orifice was increased to seven-eighths of an inch. There was no apparent regurgitation.

A lung biopsy was taken and showed moderate haemosiderosis.

Convalescence was smooth and 14 days after the operation, the child was running about the corridors with other children, and was free of dyspnoea.

The improvement was subsequently maintained.

Case 2. P.M., a 14-year-old Mosotho male, was admitted to Baragwanath Hospital on 30 July 1953 under Mr. L. Fatti. He had become ill in February 1952 and was in bed for a month without any medical attention. After this he was well until 3 weeks before admission. He attended school and played football but found that he was somewhat breathless while dribbling the ball. Three weeks before admission he noticed severe breathlessness on effort. This became worse and at the time of admission he could barely walk two blocks. Exercise produced frontal headache, giddiness, and a feeling of faintness. Exertion also caused praecordial pain. He did not complain of cough and had never suffered from haemoptysis.

On examination he was small for his age and very thin. He was not pyrexial. The radial pulse was small. There was no cyanosis.

The jugular venous pressure was normal.

The maximal cardiac impulse was in the 6th intercostal space in the mid-clavicular line, and appeared to be thrusting. There was a marked left parasternal heave. On auscultation at the apex the 1st sound was loud, there was an opening snap of the mitral valve, and a loud mid-diastolic and presystolic murmur was heard. The pulmonary 2nd sound was accentuated and normally split.

The liver was not palpable and no adventitious sounds were heard at the lung bases. There was no ankle oedema.

Radioscopy (Fig. 3). The heart was not enlarged. The cardiothoracic ratio was 49·5%. In the postero-anterior view the pulmonary-artery segment was prominent, and the left atrial appendage was visible on the left heart border. A double density of an enlarged left auricle was seen and confirmed by an indentation in the barium-filled oesophagus in the right oblique view. The pulmonary vessels especially at the bases were attenuated. There were early signs of pulmonary haemosiderosis and horizontal linear shadows as described by Kerley[3] were visible in the right costophrenic angle.

The electrocardiogram showed right axis deviation in the standard leads with a mitral P wave. There was considerable right ventricular preponderance best seen in lead AVR and lead V_1. (Fig. 4).

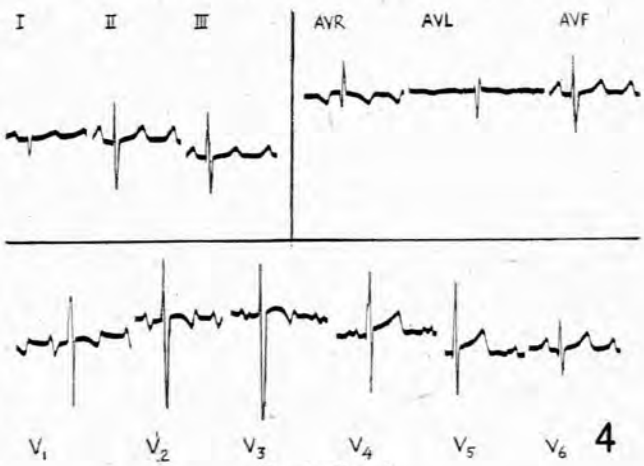

Fig. 4. Case 2.

Blood cultures yielded no growth.

At operation on 26 August 1953 (Mr. v. d. Spuy) the left atrium was much enlarged. The mitral valve was funnel-shaped and elastic. The orifice was three-sixteenths of an inch in diameter. The anterior cusp was long and mobile, and the posterior cusp was short and thick. There was no regurgitation. The valve split easily and the orifice was seven-eighths of an inch in diameter after

splitting both commissures. Before splitting the valve a diastolic thrill was felt over the left ventricle. The lungs were tense and a lung biopsy was taken.

After the operation the boy appeared well, but was still running a temperature ranging between 100° and 103° F on 4 September. This was thought to be due to a small left-sided pleural effusion, but the report of the lung biopsy received on 5 September stated there was patchy atalectasis, slight haemosiderosis, and one or two small granulomatous foci consisting of epithelioid cells and Langhan's giant cells and surrounded by lymphocytes and plasma cells. Acid-fast and alcohol-fast bacilli were not demonstrated in stained preparations, but the histological features suggested tuberculosis (Dr. W. J. Pepler, South African Institute of Medical Research). In view of this report the patient was treated with streptomycin, ½ g. daily, and Rimifon, and by 7 September the temperature was normal and remained so until 26 September when the patient was transferred to another hospital.

After 6 weeks in that hospital he was discharged, and 3 months later was free of dyspnoea and had gained weight, and his mother reported that he had grown a lot. Unfortunately he has been lost sight of since.

Case 3. This 14-year-old Xosa girl was admitted to Baragwanath Hospital under Dr. V. Wilson on 4 May 1954. Her main complaint was pain in the left praecordium which occurred after exercise. It was relieved by rest. For one year she had been dyspnoeic on mild effort and after exercise occasionally got an attack of coughing productive of white frothy sputum, which was occasionally blood-tinged. She sometimes experienced a pain in the right hypochondrium. There was no history of joint pain or swelling.

On examination she was not pyrexial. The jugular venous pressure was normal. The radial pulse was regular and full. The blood pressure was 140 mm. Hg. systolic and 75 mm. diastolic.

The maximal cardiac impulse was palpable in the 5th interspace in the mid-clavicular line. A diastolic thrill was palpable at the apex. There was a well-marked left parasternal lift. On auscultation the apical 1st sound was loud. There was a mid-diastolic rumbling murmur with presystolic accentuation. An opening snap of the mitral valve was not audible. The pulmonary 2nd sound was accentuated.

The liver was not palpable and there were no adventitious sounds at the lung bases. There was no ankle oedema. A few small enlarged lymph nodes were palpable in the axilla.

Radioscopy showed prominent pulmonary arteries in the hilar region. The heart did not appear to be enlarged. The left auricular appendage was visible on the left heart border. The left auricle was enlarged in the left oblique view and indented the barium-filled oesophagus in the right oblique view. The right ventricle was obviously enlarged in the left oblique view.

The electrocardiograph showed a mitral P wave in leads 1 and 2. The P wave in lead VI was large and diphasic. There was no evidence of ventricular hypertrophy. (Fig. 5).

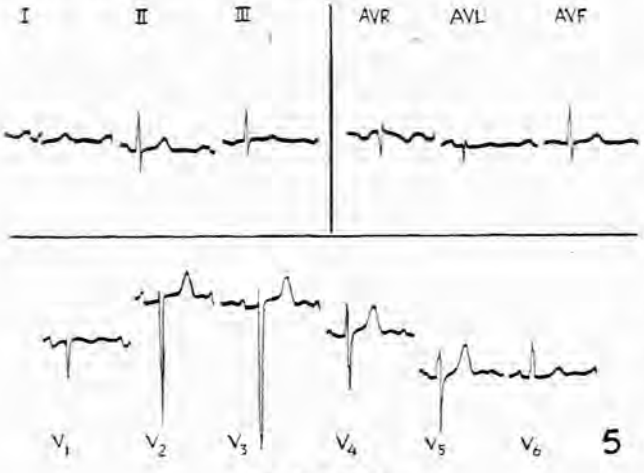

Fig. 5. Case 3.

A full blood count on 5 May was normal, the E.S.R. was 11 mm. in 1 hour (Westergren). The C-reactive protein was 2+ positive.

Operation. On 19 May mitral valvotomy was performed by Mr. L. Fatti. The left atrium was considerably enlarged. The left ventricle appeared normal. The left auricle was entered without difficulty through the left atrial appendage. The mitral valve was tightly stenosed and diaphragmatic in type, the orifice being three-eighths of an inch in diameter, and the cusps were elastic and billowing. A valvotome was used to cut the lateral commisure and for blunt dissection to open the medial commissure. The final orifice of the mitral valve was 1 inch in diameter and there was no regurgitation. A well-marked diastolic apical thrill on the left ventricle disappeared at the end of operation.

The post-operative course was smooth apart from moderate pyrexia on the first 3 post-operative days. On discharge on 15 June 1954 no mid-diastolic murmur was audible and effort tolerance was good.

Case 4. This 14-year-old European boy was admitted to a private hospital under Mr. L. Fatti on 30 January 1954.

He had suffered an attack of rheumatic fever at the age of 4 years, and had been breathless since then. He had been admitted to Pietersburg Hospital several times for acute cardiac failure, and twice (1952 and 1953) was thought to have pulmonary infarction. On one occasion in 1952 he was pyrexial, and although blood cultures were negative he was treated as a subacute bacterial endocarditis. He contracted frequent attacks of bronchitis in the winter, and during these attacks became very short of breath.

On admission he stated that he could only walk 100 yards on the level, and could barely climb one flight of stairs. He slept on three pillows. On walking he complained of dull substernal pain. This did not radiate, but it caused him to stop walking until it passed off—usually in about 1 minute. In 1948 he had oedema of the legs but this had not recurred since. In January 1953 he developed acute appendicitis, which was treated conservatively because of his heart condition. This settled down without any further trouble.

He was taking digoxin, 0·25 mg. twice daily, and 0·5 g. of sulphatriad daily, but had not had any mercurial diuretic injections for some months before admission.

On examination at admission there was slight peripheral cyanosis of the fingers and toes. There was deep jugular venous pulsation, mainly systolic, suggesting tricusped incompetence. The jugular venous pressure was normal. The radial pulse was regular and slightly collapsing. The blood pressure was 110 mm. Hg. systolic and 70 mm. diastolic.

There was a praecordial 'bulge'. A diastolic shock was palpable over the pulmonary artery. There was a marked left parasternal lift, and the apical impulse was a diffuse heave in the axilla suggesting right ventricular enlargement. On auscultation at the apex a scratchy grade-2 systolic murmur was heard. This varied in intensity from day to day. A grade-3 mid-diastolic and presystolic rumbling murmur was audible in the axilla, and the 1st sound at the apex was loud and snapping. Just medial to the apex an opening snap of the mitral valve was audible but soft. The 2nd sound at the 2nd left intercostal space was loud and normally split, the 2nd component being accentuated. A grade-2 early diastolic murmur was present at the left sternal border.

The chest was barrel-shaped. A few high-pitched rhonchi were heard at the bases. The liver edge was palpable 2 finger-breadths below the costal margin and at times systolic pulsation was present. The spleen was palpable 2 finger-breadths below the costal margin.

An electrocardiogram showed a severe degree of right ventricular hypertrophy (Fig. 6).

Laboratory investigations including serum muco-proteins, full blood-count, C-reactive proteins and several E.S.R. estimations gave normal results.

X-ray showed a small aortic knuckle, prominent pulmonary-artery segment, an enlarged left auricle and right ventricle, and considerable cardiac enlargement. The lung fields showed haemosiderosis, peripheral attenuation of the pulmonary vasculature and some Kerley B-lines in the right costophrenic angle suggesting

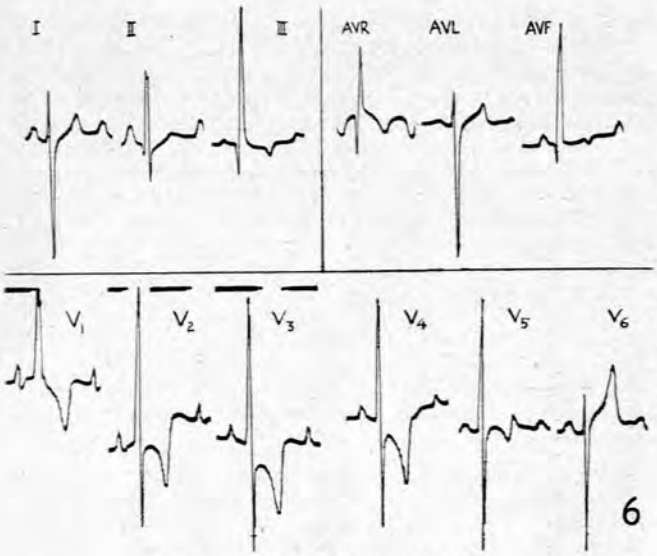

Fig. 6. Case 4: 25 February 1954.

severe pulmonary hypertension. Serial X-rays over 6 years showed progressive cardiac enlargement. (Figs. 7 and 8).

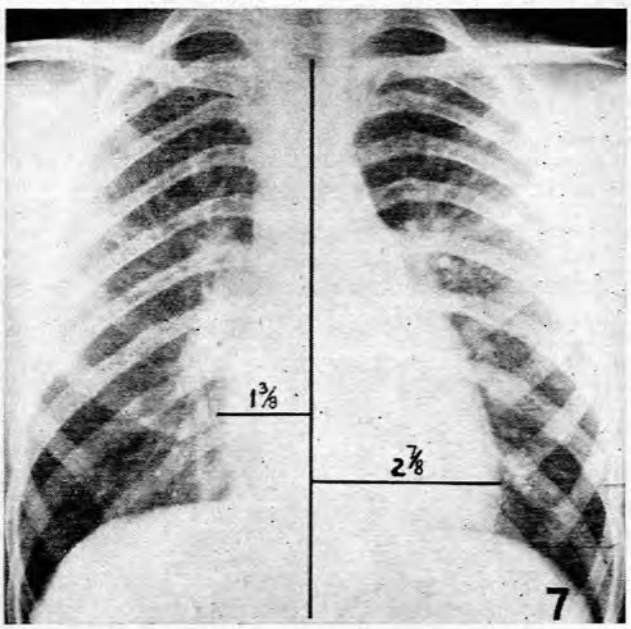

Fig. 7. Case 4: 16 October 1948. Figs. 7 and 8 show the progressive changes in size and shape of heart.

Operation was performed on 12 May by Mr. G. R. Crawshaw. The approach was through the 4th left intercostal space. The left atrium was large and tense. Both the mitral cusps were soft and mobile. The valve orifice was postero-lateral in position and oval in shape, being ¾ inch by one-eighth of an inch. The valve split easily along the commissures with finger fracture and the post-operative size was 1¾ by 1 inch, with no regurgitation. The right ventricle was seen to be very large and the lungs were stiff and showed pigmented subpleural spots, sparing the apices. A lung biopsy showed haemosiderosis and early endarteritis in the arterioles. A section of the atrial appendix showed slight perivascular fibrosis but no evidence of active rheumatic disease.

The post-operative course was smooth, but was interrupted on the 15th post-operative day by an attack of acute appendicitis.

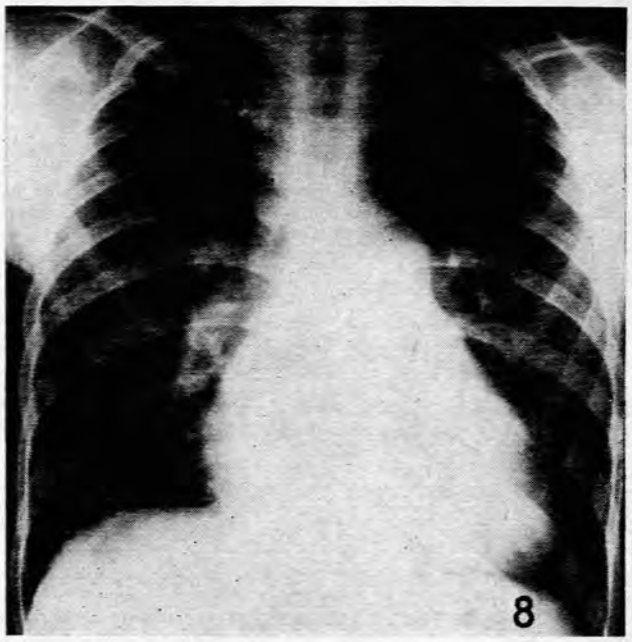

Fig. 8. Case 4: 4 March 1954. See Fig. 7.

This was dealt with by operation (Mr. R. Frylinck) and subsequent recovery was uneventful. The boy's exercise tolerance improved dramatically and 10 days after his appendicectomy he walked a quarter-mile without any fatigue. A grade-1 mid-diastolic rumble was audible at the cardiac apex but there was no evidence of an opening snap.

He was discharged on a maintenance dose of digoxin and oral penicillin (Bicillin), the latter to be continued for 2 years.

On 28 August 1954 Dr. F. A. B. Lombard reported from Pietersburg that the patient had had 3 severe pyrexial attacks—2 in April and 1 in July—during which he complained of severe pain in the left side of the chest and dyspnoea. Tachycardia and cyanosis were present. In the attacks in April there were no chest signs. He was treated with large doses of antibiotics and recovery occurred in about 48 hours each time. In July clinical examination and radiography showed consolidation in the left upper lobe. Terramycin was administered and he responded well, being pyrexial for only 3 days. Apart from these attacks he was well and was riding his bicycle. He had gained 25 lbs. in weight and his height had increased by 4½ inches since the operation. Dr. Lombard remarked

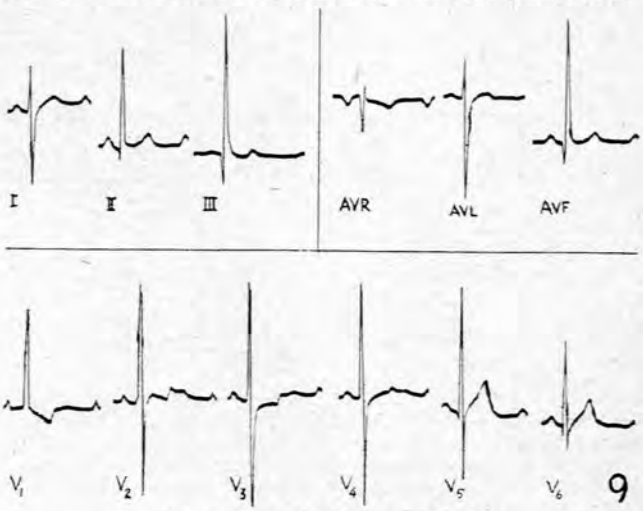

Fig. 9. Case 4: 18 August 1954.

that 'there is no doubt that this young man's whole life—mentally and physically—has been improved by operation'. The electrocardiogram (Fig. 9) showed considerable improvement. An X-ray showed no material change in heart size. The haemosiderosis was still present.

Comment. This case shows several interesting features. In general the occurrence of anginal pain in mitral stenosis is not uncommon, but at this age it must be unusual.[4] The splenomegaly may have been due to some associated condition such as bilharziasis in view of his residence in the North-Eastern Transvaal, but splenomegaly can occur in long-standing congestive failure. There is no evidence of subacute endocarditis. The post-operative pyrexial incidents in April are of the type known to occur after mitral valvotomy in 10-20% of patients.[7,9] Its aetiology is unknown, and has been explained on the basis of re-activation of rheumatic fever, or of pericardial foreign-body reactions.

DISCUSSION

These 4 cases show various features of interest. They all presented indications for valvotomy. Two (cases 1 and 4) were in congestive cardiac failure due to high pulmonary vascular resistance,[9] while 2 had attacks of paroxysmal dyspnoea or pulmonary oedema on effort (cases 2 and 3). It is extremely unlikely that any of these patients would have survived very long without valvotomy, and all showed considerable subjective and objective improvement after operation.

It is usually said that mitral valvotomy should not be performed below the age of 17 years, and that great caution should be exercised up to the age of 20 years, because active or latent rheumatic carditis increases the risks of operation and the rheumatic state may be re-activated; furthermore, recurrences of rheumatic fever are more common in adolescence and may cause further valvular damage in the years after operation. However, it is agreed that if the mitral stenosis is severe enough to threaten life, operation should not be withheld.[1] There is no real evidence that operation re-activates a rheumatic process,[4] and mitral stenosis in itself may be a more dangerous condition than the rheumatic fever. While overt acute rheumatic fever would contra-indicate operation, a latent or doubtful state of rheumatic activity should not interfere with the decision to operate if real indications are present. Should rheumatic fever recur, with recurrence of mitral stenosis, a further valvotomy can, in many cases, be performed.

SUMMARY

(1) Mitral stenosis is rare under the age of 16 years.

(2) If the mitral stenosis threatens life, valvotomy should be performed whatever the age.

(3) Doubtful or latent rheumatic fever is not a contra-indication to operation.

(4) Four cases, aged 14 and under, who have undergone successful valvotomy are described.

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