Extreme pulmonary hypertension caused by mitral valve disease
Natural history and results of surgery

C. Ward and B. W. Hancock

From the Cardio-Thoracic Unit, Northern General Hospital, Sheffield

Five hundred and eighty six patients with mitral valve disease were studied with cardiac catheterization between 1961 and 1972; 48 (8.2%) had extreme pulmonary hypertension (resting systolic pulmonary artery pressure of 80 mmHg or above and pulmonary vascular resistance of 10 units or greater) and of these patients, 27 underwent cardiac surgery. The operative mortality for mitral valvotomy was 11 per cent and for mitral valve replacement 56 per cent. The overall mortality was 31 per cent. The risks of operation were increased in those with a long history of cardiac symptoms, those over 50 years of age, and in the presence of associated aortic valve disease. The mean survival for those patients not having operation was only 2.4 ± 0.5 years. The mean follow-up period for those surviving operation has been 5.8 ± 0.6 years, and symptomatic improvement has been good.

The operations of mitral valvotomy and mitral valve replacement have greatly improved the prognosis of patients with mitral valve disease. It is generally recognized that patients with severe pulmonary hypertension are less likely to survive such operations (Emanuel and Ross, 1967). Despite this, it has been suggested, because of the potential benefits, that patients in this situation should not be refused surgery (Hamer et al., 1968). Barclay et al. (1972) considered a systolic pulmonary artery pressure of 110 mmHg or greater to be an absolute contraindication to mitral valve replacement. The increased risk of operation in these patients can only be justified if the prognosis is thereby improved. We have, however, been unable to find any report which specifically outlines the natural history of extreme pulmonary hypertension caused by mitral valve disease and reports of mitral valve surgery on such patients do not indicate the expected course of the disease in a comparable group not having surgery. The objectives of this study have been to outline the natural history of extreme pulmonary hypertension secondary to mitral valve disease, to determine the risks of surgery in such patients, and to discover if there is a group of patients in whom the risks of operation are not justified.

Patients and methods

Case records from the Cardio-Thoracic Unit of the Northern General Hospital, Sheffield have been examined for the years 1961 to 1972. Data on 586 patients with a principal diagnosis of mitral valve disease, who had had cardiac catheterization, were studied. Forty-eight patients had extreme pulmonary hypertension defined arbitrarily as a resting pulmonary artery systolic pressure equal to, or greater than, 80 mmHg. Details of the following were noted: pulmonary artery pressure; wedged pulmonary artery pressure (indirect left atrial pressure); electrocardiographic evidence of atrial fibrillation and of left and/or right ventricular hypertrophy; x-ray evidence of cardiomegaly, left atrial enlargement, and pulmonary hypertension; presence of mitral valve calcification; relevant medical history; and the nature of valve lesions. An estimate of pulmonary vascular resistance was made on the basis of Wood's findings (1954), who showed that it was linearly related to the pulmonary pressure gradient (mean pulmonary artery pressure – mean left atrial pressure); a pulmonary vascular resistance of 6 to 10 units occurring in those patients with a pulmonary pressure gradient of 15 to 30 mm and a pulmonary vascular resistance of 10 to 30 units with a pulmonary pressure gradient of 30 to 70 mm.

The New York Heart Association functional classification has been used in the clinical assessment of the patients.

Cardiac catheterization had been performed on each patient as a preliminary to consideration for surgery.

Received 28 May 1974.
Those patients who were not operated on after cardiac catheterization were classed as the 'nonoperated' group (group 1). Those who underwent operation were classed as the 'surgical' group (group 2). The latter (group 2) was further subdivided into those who survived the postoperative period (group 2A) and those who did not (group 2B).

The findings in these groups are summarized in Tables 1 and 2. Statistical significance has been assessed from values for probability (P) based on Students' t-test or $\chi^2$ test.

**Results**

**All patients**

Of the total of 48 patients, 15 had a pulmonary arterial systolic pressure of 80 to 89 mmHg, 9 of 90 to 99 mmHg, 12 of 10 to 109 mmHg, and 12 of greater than 110 mmHg. In 3 patients, the pulmonary arterial pressure was greater than 150 mmHg. The pulmonary vascular resistance was 10 to 15 units in 26 patients (54%) and greater than 15 units in 22 patients (46%).

**Group 1: no operation**

This group consisted of 21 patients (12 female and 9 male). At the time of catheterization the mean age was $46.2 \pm 1.7$ years and the length of history $7.4 \pm 1.1$ years. At that time 1 patient was in functional class II, 14 were in class III, and 6 in class IV. Seven patients had additional significant aortic valve disease. Only 6 patients had isolated mitral stenosis. The remainder had additional mitral regurgitation and/or aortic valve disease. Two patients have survived (both in functional class IV) one for 6 years and the other for 4 years.

**Group 2: all surgical patients**

In all, 27 patients (19 women and 8 men) underwent 29 operations. At the time of catheterization the mean age was $42.4 \pm 1.6$ years and length of history $5.6 \pm 1.3$ years. Two patients were in functional class II, 19 in class III, and 6 in class IV. Eighteen patients had mitral valvotomy (1 with additional aortic valve replacement). Nine had mitral valve replacement and 2 mitral plus aortic valve replacement.

**Group 2A: patients surviving surgery**

Nineteen patients (16 female and 3 male) survived 20 operations. The mean age at the time of cardiac catheterization was $40.4 \pm 1.3$ years. At this stage,

<table>
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<th>Table 1 Comparison of results in nonoperated and surgical groups</th>
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<td><strong>Group 1</strong></td>
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<td><strong>Patients not having surgery (21 patients)</strong></td>
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<td>Mean age (± SE mean) at time of catheter assessment (yr)</td>
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AF = Atrial fibrillation.
1 patient was in functional class II, 13 in class III, and 5 in class IV. The length of history was 4.2 ± 0.8 years. The operation was mitral valvotomy in 15 patients, mitral valvotomy plus aortic valve replacement in 1, and mitral valve replacement in 4. Fifteen patients are alive at the time of writing (mean follow-up period 5.8 ± 0.6 years). There have been 4 late deaths — 1 from bronchiolitis, 1 after a road traffic accident, 1 with congestive cardiac failure 6 years after operation, and 1 after further cardiac surgery. Of the survivors, 3 are in functional class I, 11 in class II, and 1 in class III.

**Group 2B: patients dying in postoperative period**

There were 9 postoperative deaths (3 female and 6 male). The mean age at the time of cardiac catheterization was 47.1 ± 3.8 years and length of history 9.3 ± 2.1 years. At this stage 1 patient was in functional class II, 7 in class III, and 1 in class IV. Of the 6 patients aged 50 years and over at the time of surgery, 5 died postoperatively. Of the 9 patients, 5 had mitral valve replacement and 2 mitral valvotomy. Both patients who had mitral plus aortic valve replacement died after operation. The incidence of mitral valve calcification and atrial fibrillation was significantly increased in this group compared with the surgical survivor group.

**Discussion**

Extreme pulmonary hypertension secondary to mitral valve disease has a poor prognosis, and though surgery is more hazardous in these patients than in those with lower pulmonary artery pressures, the potential benefits appear to justify the risks in the majority of patients.

Olesen (1962) has shown in a purely clinical study that the prognosis for medically treated patients with mitral valve disease is worse when there is clinical evidence of increasing pulmonary hypertension. Ten (48%) of 21 patients in our series who did not have surgery died within a year of cardiac catheterization. This group, however, is not strictly comparable with the surgical group because of the more common additional presence of aortic valve disease. The mean survival of those patients with associated aortic valve disease was 1.5 ± 0.9 years. Even without this high risk group, the mean survival of the remaining 14 patients was only 2.9 ± 0.6 years. The reasons for not operating on patients after cardiac catheterization were not...
always made clear in the case records but may be
surmised. Many of the patients were from the
earlier years of the study when surgeons were less
inclined to operate on high risk patients, particu-
larly as many of these patients would have needed
mitral valve or double valve replacement operations
which were not then being routinely performed.

Mitrval valvotomy and mitral valve replacement
have dramatically improved the outlook for many
patients with mitral valve disease (Emanuel,
1963; Hamer et al., 1968). Nichols et al. (1964)
reported on an overall surgical mortality for all
types of mitral valve surgery, of 8 per cent increas-
ing to 23 per cent for those with a pulmonary arterial
pressure greater than 50 mmHg. Hamer et al.
(1968), reporting on their earliest mitral valve re-
placements, noted a mortality of 25 per cent which
rose to 35 per cent in patients with a pulmonary
vascular resistance greater than 10 units. Three out
of four of their patients with a pulmonary vascular
resistance in excess of 14 units died after operation.

It has been suggested that other factors increase
the hazards of operation (Turner, 1967). In the
present study, atrial fibrillation, mitral valve calci-
fication, and a long history of symptoms were more
common in those who died after operation than in
those who survived. These patients were also older
than the survivors. The situation is complicated by
the frequent occurrence of these factors in com-
bination in the same patient, making it difficult to
assess the part played by any one of them in affect-
ing the prognosis. Emanuel (1963) has helped to
clarify the situation. The mortality for his patients
undergoing mitral valvotomy who had a pulmonary
vascular resistance of 10 units or greater was 13 per
cent, this figure rising to 26 per cent if there was
mitral valve calcification and to 50 per cent with the
additional presence of mitral regurgitation. Most
reported series relate mortality primarily to the
level of pulmonary hypertension, and in interpreting
results it must be remembered that other factors are
contributing, though to what extent is not always
clear.

Patients in this study who underwent surgery, all
had a pulmonary vascular resistance of greater than
10 units, and in 13 it was greater than 15 units. The
overall operative mortality was 31 per cent, 11 per
cent for mitral valvotomy and 56 per cent for mitral
valve replacement. Five patients died after mitral
valve replacement. The pulmonary arterial pressure
and pulmonary vascular resistance in these patients
was not significantly different from that recorded in
the survivors. It was noted that no patient over the
age of 45 years survived mitral valve replacement;
the duration of symptoms in those patients who
died after operation was longer than in the sur-
vivors (mean 9.3 ± 2.1 years, as opposed to 42 ± 0.8
years) and men were less likely to survive than
women. The 4 who had survived mitral valve re-
placement are alive 1½, 2, 2½, and 4 years, respec-
tively, after operation. The mean survival for all
surgical patients surviving operation has been
5.8 ± 0.6 years. For the medically treated group,
even excluding those with additional aortic valve
disease, the mean survival has been only 2.9 ± 0.6
years. Symptomatic improvement in those who sur-
vive operation is also obvious. At the time of cardiac
catheterization, 31 patients were in functional class
III and 12 in class IV. The 2 patients in the medi-
cally treated group, alive at the time of writing 4 and
6 years after investigations, are in functional class
IV. The other members of this group remained
severely incapacitated until they died. Fifteen
patients from the surgical group are alive, 14 of
these being in functional classes I and II. The 4
patients who survived mitral valve replacement were
in class II when last seen.

The poor prognosis for patients who do not have
operation has not previously been sufficiently
stressed. A quarter of the patients in our study died
within 6 months of cardiac catheterization and a half
within 12 months.

Extreme pulmonary hypertension has been con-
sidered a relative, if not absolute, contraindi-
cation to valve surgery particularly in those requiring
mitral valve replacement. The findings presented
here suggest that the reverse should be the case.
Extreme pulmonary hypertension should be re-
garded as a positive indication for early operation
in many of these patients. No patient should be refused
mitral valvotomy because of extreme pulmonary
hypertension, neither should most patients under
the age of perhaps 50 to 55 years be refused mitral
valve replacement. However, patients older than
this, those with a very long history of breathlessness,
and those needing additional aortic valve replace-
ment remain a serious problem and surgery still
has a very high mortality in such patients. It is
likely that a group of such patients exists in whom
the risks of operation are still not justifiable. It is
anticipated that this group will steadily diminish as
postoperative care improves.

We are grateful to the physicians and surgeons of the
Cardio-Thoracic Unit, Sheffield, for allowing us to
report details of the patients and to Professor A. M.
Walker for help with statistical analysis.

References

Barclay, R. S., Reid, J. M., Stevenson, J. G., Welsh, T. M.,
valve replacement with Starr-Edwards prostheses.
British Heart Journal, 34, 129.


Requests for reprints to Dr. C. Ward, Northern General Hospital, Sheffield S5 7AU.
Extreme pulmonary hypertension caused by mitral valve disease. Natural history and results of surgery.
C Ward and B W Hancock

Br Heart J 1975 37: 74-78
doi: 10.1136/hrt.37.1.74

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